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Thomas Fairbank

THOMAS FAIRBANK BIRTHDAY VOLUME

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EVERY CONTRIBUTION IN THIS NUMBER HAS BEEN SPECIALLY INVITED
IN HONOUR OF THE EIGHTIETH BIRTHDAY OF SIR THOMAS FAIRBANK

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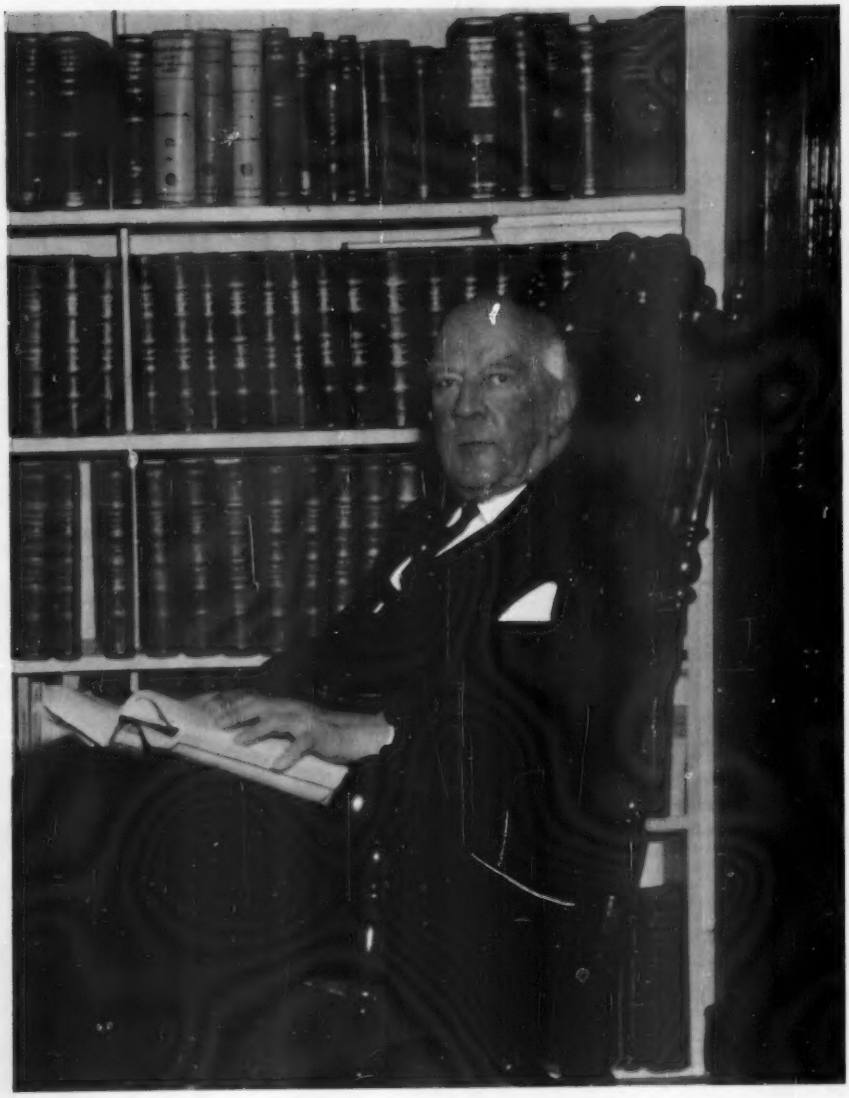


The Journal of Bone and Joint Surgery

THOMAS FAIRBANK BIRTHDAY VOLUME

" I will never be an old man; to me old age is always fifteen years more than I am." So it is with Thomas Fairbank who will never be old, not even within the next fifteen years. The vivacity of his spirit and the industry of his activity have kept him young all the time. It is true that this special issue of the British volume of the *Journal of Bone and Joint Surgery* is a Birthday Volume commemorating eighty years of vigorous endeavour, in which every invited contributor is proud to pay respect, regard and warm affection to " Uncle Tom "; but all who feel so proud of him, and are so glad to have known him and been stimulated by him, expect very many more years of his friendship.

The great contribution that he has made to the advance of orthopaedic surgery through teachings and publications is acknowledged in these pages; but we must also emphasise the qualities of kindness and understanding that have made him so great a leader of young surgeons both in days of peace and days of war. Such quiet and mellowing influence is not always recognised but it represents a most important part of the life of our doyen of British orthopaedics.

There are many who have worked long and faithfully with Sir Thomas Fairbank, including especially his operating sister, private secretary and librarian, Miss B. Florence Bradbury. But a very special tribute is owed in this Birthday Volume to Lady Fairbank. Every single one of us who has known her will join in an expression of love to one who has so loved a great man that she too has made the one gift that matters in this world—the relief of suffering and anxiety and the conferment of happiness and joy upon those who need it—and who does not?

REGINALD WATSON-JONES

AN OPEN LETTER

FROM SIR ROBERT HUTCHISON, BART.

MY DEAR FAIRBANK,

So you have joined the ranks of the octogenarians! Being myself a member of that select body I should like to offer you a cordial welcome to it—*Finis coronat opus*, and I think you will find that to have exceeded the three score and ten by a decade has its compensations. It must be fully half a century since we first were colleagues at Great Ormond Street soon after your return from the South African War, and, although our paths have diverged since then and of late years I have not seen as much of you as I could have wished, yet I have followed your career with interest and admiration and have been glad to note how the promise of your early days at the Hospital for Sick Children has been so amply fulfilled. Others, better qualified than I, will describe the great work you have done in your special branch of surgery, but I should like to say, if I may, how much all of us have been inspired by the high standard of professional integrity of which you have always set an example. I join therefore most cordially in the acclamations which will greet you on your eightieth anniversary and in the hope that you will have many years yet of health and happiness in which to enjoy the high reputation and esteem you have so richly earned.

I am, my dear Fairbank, with every good wish,

Yours very sincerely,

ROBERT HUTCHISON

SIR THOMAS FAIRBANK

AN APPRECIATION BY ST J. D. BUXTON

Fairbank is a man of outstanding character and brilliance. During his long and active career he has been an eminent orthopaedic surgeon. No one can write about his life and work without realising his charm and great courtesy, so evident when he is approached on a scientific problem, when a host to a few, or at an association function. He has been able to help many, because all who have met him quickly appreciate the great knowledge, the sincerity, the analytical mind and the breadth of vision that he possesses. His talent and personal qualities became more evident as friendship developed.

It is a welcome occasion to one who has had the good fortune to enjoy Tom Fairbank's friendship and had the opportunity of working under and with him almost since the day of qualification to reflect on these years, but it is not an easy task to assemble and assess the evidence in order to express the true value and influence of Fairbank's life and work. His association with famous institutions and public services has been important; hence it appears that a study of his work and writings during periods of his life make it possible to realise the part he played in the development and advance of knowledge of orthopaedic surgery in the first half of this century.

EARLY YEARS

He is one of a family of five, whose father Dr Thomas Fairbank died when the children were small. Dr Fairbank was in practice in Windsor and in the 1870s was doctor to the Castle and to Princess Christian and her family. " H. A. T." was educated at " Epsom " and qualified at Charing Cross. Before the century ended he was also a qualified dental surgeon and on his way to South Africa with " the gentlemen in khaki ordered south." He volunteered as a civil surgeon, as he thought the experience would be of benefit in gaining a knowledge of the world as well as providing an opportunity of learning medicine and surgery of a type not reaching a teaching hospital. It was possible to obtain an appointment for one year and travel out with a general hospital, the staff being augmented by the volunteer civil surgeons. The study of family records during this experience is not only of interest, but reveals his enthusiasm, powers of observation and description.

South Africa is very different to-day, and he was able to see Cape Town, Johannesburg, then only fifteen years old, and Pretoria, then a small town. During the campaign he met Lord Roberts and Lord Kitchener, the first time being when they visited a hospital in the middle of the night in mufti. Kipling and Conan Doyle visited a hospital where he worked. For some period he was at De Aar, living in a tent in " Harley Street," and was employed doing some surgery which was particularly welcome and in charge of many sick with typhoid fever. He records that on January 27, 1900, he had his first experience of removing a bullet.

He was glad to be on trek, usually riding, and was interested in the fighting area, recording his reactions and the taking of a photograph of a gun under fire. He was at Roberts's camp at Paardeberg and saw the surrender of Cronje, riding a white horse into our line. He stated that the iron ration of the day was made up in an iron cylinder, one part containing cocoa and the other compressed beef and was supposed to be sufficient to sustain the body for thirty-six hours.

It would seem that there were few dull moments and certainly amusing episodes, such as the visits of Lady Roberts to the hospitals, in which she was interested and undoubtedly helpful.

Before the year ended he realised he wanted to return to continue studies and obtain higher qualifications and he was able to do this, having earned the South African medal with five bars.

LONDON 1900–1914

Two years' work as senior resident at the Hospital for Sick Children, Great Ormond Street, was soon followed by staff appointments at that hospital and at Charing Cross Hospital so that he was able to study, develop powers of observation and gain experience in diagnosis and operative treatment. He was introduced to surgery particularly by Arbuthnot Lane and Boyd, and during subsequent years there is no doubt that friendship amongst colleagues at Ormond Street—George Waugh, Oswald Addison and Tyrrell Gray—encouraged each and all in surgical planning and clinical observation at a high level. Lane was developing his "no-touch" technique, and gloves—at first heavy and wet—were coming into use. Caps came a little later. Fairbank was interested and practised the Lane technique most meticulously all his career.

At the Hospital for Sick Children, about 1909.

Orthopaedic surgery came to him early and he met Lorenz when he visited London in 1903. During the first decade of this century, when few surgeons in England took any particular interest in this subject, Fairbank wrote papers on Sprengel's shoulder, talipes, coxa valga, congenital dislocation of the hip and in 1910 reported three cases treated by Lorenz operations.

During these years he began observations on the affections of the skeleton in children and acquired the friendship and admiration of many child patients and their parents. It appears he made observations of value on other surgical conditions, and over fifty years ago he wrote on intubation of the larynx and has recently pointed out to me renewed interest in this subject. The staff appointment at Charing Cross gave less opportunity to him than at Ormond Street, for the time had not arrived when teachers of general surgery wished the students to receive regular instruction from more than a few special departments and few beds were available for specialists. During this period some hours were spent with a Territorial Field Ambulance; so he was prepared when the crisis of the first world war arose in August 1914. Earlier that year he visited the States, going to New York, Boston, Philadelphia, Portland, Maine, where he met Abbott, Cleveland to see Murphy, and to stay a short time at Rochester where the Mayo Clinic was well established. In later years he probably talked more of Osgood and Abbott than of others he met. By that time Boston had developed a large orthopaedic clinic.

THE FIRST WORLD WAR

Fairbank was already commissioned in the Royal Army Medical Corps, Territorial Force. The regulations on the outbreak allowed for little opportunity for a surgeon attached to a unit unlikely to require top level specialists, to be suitably employed; so he and other London consultants remained in the Field Ambulance for some years. Details of his service in France and Macedonia are described elsewhere (page 22) and it will be noted that, when the decision was made to transfer territorial officers to posts in which their services were particularly valuable, Fairbank lived at a general hospital at the Base (Salonika area) and worked with Robert Kelly (Liverpool). Crisp English (St George's) had been Senior Consultant Surgeon, and when he moved to the Italian area, Kelly took his place. Now Fairbank was able to travel to the Doiran and Struma areas to help the younger surgeons at casualty clearing stations and hospitals on the lines of communication. The travelling was tiring in those days as motor cars for rough roads were almost in the " veteran " class. No saloon car with large tyres was available and he traversed mountain tracks in a car with a high body, inefficient hood and poor springs. In the summer the heat was considerable and in the winter there was little protection from Macedonian storms. However, he never put off an arranged visit and was always up to time. In the tented wards and in the mess he was most helpful and his initiative in tackling routine or special problems encouraged those he had come to help. Naturally the problem of the infected fracture was foremost. Could the infection be prevented? How should the limb be immobilised? Was " Carrel Dakin " the best method of treatment, or would acriflavine as an irrigation or dressing take its place? For the surgeon in the forward area he was most helpful in providing interchange of information from other areas, for surgical meetings were difficult to arrange, and for reporting progress of patients passed on. His assistance in decisions regarding movement of patients down the line were particularly useful, for roads were shocking and transport limited. On these expeditions from the Base the limited kit always included the net and paraphernalia for catching and setting butterflies, for the thistles on the hills were the habitat of Camberwell Beauties, and heaths and blues were plentiful, and swallow tails were caught. The half-hour after tea to " see what I can get " extended to an hour and a half hunt in the nullahs, which was carried out with the same enthusiasm and method as the work in the hospital.

An important problem tackled by Fairbank was at the convalescent depot at Hortiach, a few miles from Salonika. Sir Percy Holt, the Director of Medical Services, had to deal with the serious wastage of personnel due to sickness in the Salonika Force of about 100,000. This was due particularly to malaria and dysentery, but it was found that there was an accumulation of officers and men who had been discharged from hospital after some treatment for minor injury. It was routine to send them to a convalescent depot and then to a regimental depot. Little or no treatment was available and the soldier was often "browned off" with recurrent attacks of fever, short stays in several hospitals in an area where he had no entertainment or hope of seeing " blighty " for some time. Fairbank saw the problem, which included a far too large assortment of undiagnosed and untreated conditions of the knee and other joints. In a short time those in the depot were sorted out and a unit working, which relieved physical ailments and restored morale. He set Marlin, who subsequently specialised in physical medicine, on to developing a convalescent depot, to which every surgeon wished to send his patients. This was undoubtedly the first rehabilitation centre overseas organised on principles employed today. These principles although employed at home in the organisation set up by Robert Jones were almost forgotten during the 1930s. Perhaps this was the most useful work Fairbank did in Macedonia. It never received general recognition and many would remember the personal contact with Fairbank and forget what he had organised for the after-treatment of their patients.

Three years of Macedonia under the prevalent conditions tired out body and mind of most, and it was fortunate that the end of 1918 saw Fairbank's work there complete. Some months of well deserved leave soon saw him back at work in London.

LONDON 1919–1939

There is little doubt that Fairbank enjoyed this period of his life, during which he did such valuable work, including observations and recording of abnormalities of the skeleton to be published at a later date. It appears that his work was possible owing to his own competence and personality, to the development of the speciality of orthopaedic surgery and to the advance in diagnostic radiology.

Sir Robert Jones laid the foundation for the recognition and growth of orthopaedics in the pre-war period. He was called on to assist the medical services during the war and from that time the organisation of the speciality, the assembling of those already practising this branch of surgery and the education of orthopaedic surgeons for the future was assured. Fairbank had little contact with him until he returned from the war. Then for a short time he worked at Shepherd's Bush, one of the special hospitals, under the War Office and later under the Ministry of Pensions, which had been developed on lines advocated by Robert Jones. At each of these units the teamwork of orthopaedics was taught and employed and the *entente* between clinician and ancillary branches of surgery (physiotherapy, occupational therapy, etc.) was very close. The work at Shepherd's Bush was diminishing and Fairbank's time was required elsewhere.

The apparatus and films for radiography and the knowledge of radiologists were developing fast between 1910 and 1920. Fairbank found invaluable the association with Bertram Shires, the radiologist of the Hospital for Sick Children from 1921 until his death in 1941, and of St Thomas's Hospital. In the early 1920s he had good apparatus at his hospitals, in his consulting suite, in nursing homes and was an observer of outstanding ability. There is no doubt he helped in the differentiation of skeletal affections—particularly of children—and aided Sir Thomas in the earlier days of collecting radiographs and prints. Any surgeon who was present when the two discussed an unusual radiograph realised the rate of progress of radiology and orthopaedics in this period and that a great deal of the knowledge resulting was due in London to the collaboration of these two.

The Committee and his colleagues had missed him and his work at the Hospital for Sick Children, and his return was most welcome. During 1919 he accepted the invitation to the appointment of Senior Orthopaedic Surgeon to King's College Hospital; so he resigned from Charing Cross Hospital. King's had moved just before the war to a new building in south-east London, where hospital services were badly needed. The numerical demand by students doing their preliminary studies at King's College, Strand, was great. The Committee decided to enlarge and strengthen the staff and develop special departments. Fairbank joined the staff in company with John Thomson Walker and Kinnier Wilson, and had the support among others of Frederick Still and Frew, two colleagues at H.S.C., F. F. Burghard, Senior Surgeon, and Willoughby Lyle, the Dean. Subsequently he gave his services to Queen Alexandra's Military Hospital, Millbank, and to Lord Mayor Treloar Orthopaedic Hospital (Treloar Cripple Hospital), Alton, Hants.

Hospital for Sick Children—Fairbank was associated earlier with Thomas Barlow and Arbuthnot Lane; Archibald Garrod soon left to become Regius Professor of Medicine at Oxford, and Robert Hutchison and Still were active. The surgical staff was of the same calibre. There is little doubt about Fairbank's devoted service here until regulations necessitated his resignation as early as 1928, but important work was done in the post-war period. He demanded and established the principle that his work on the senior staff included an out-patient session each week. At that time it was customary in London for senior staff to be surgeons in charge of in-patients and for their juniors to be in charge of out-patients.

He, with F. J. Poynton, Addison and Tyrrell Gray, initiated the idea and planned a country hospital, as part of the present one in Great Ormond Street, a thickly populated area. This was made possible by a legacy of £30,000 left by the late Mr Peter Reid.

"The Country Branch, The Hospital for Sick Children, Tadworth, Surrey," was originated in 1926 as a recovery home. It developed during the second world war and is now dealing with a substantial proportion of acute medical and surgical cases. Fairbank has expressed delight at the success of this development and it is interesting to note that one-fifth of the one hundred beds are allocated to an orthopaedic surgeon, recently appointed.

Fairbank's out-patient department was filled from all over the country. He was the outstanding authority in England on the treatment of congenital dislocation of the hip and for a time the only surgeon doing "the upper lip" operation. It was evident that he had an extensive knowledge of all orthopaedic conditions of children and made progress by painstaking observation, particularly in what may be called the "difficult case." His colleagues sought every opportunity to consult him, and former residents, always capable men, wanted his opinion. Ward sisters, residents and clinical assistants enjoyed working for him. Surgeons and radiologists were sorting out the handicapped, dwarfed and disabled. Fairbank was in his element tying on the label of dyschondroplasia, marble bones or melorheostosis, or in asking Still, a great doctor and capable scholar, for a name for an undescribed condition. During these years at this hospital a large collection of the unusual was made. Notes and films were taken home and after careful scrutiny, often with friends and colleagues elsewhere, were filed for comparison with a set, which inevitably came to him later. Many of these patients were shown at the Royal Society of Medicine and their records published in the Atlas, to which reference is made later.

The early retirement necessitated many children coming to King's, but it was noticeable that his services were frequently required at the Children's Hospital and he is now a periodic visitor. His achievements there were largely clinical, but he assisted in the development of the country hospital and established the principle that any surgeon, if he wished, should have an out-patient session.

King's College Hospital—Fairbank was senior orthopaedic surgeon from 1919 to 1936. At the time of his appointment Elmslie had his department at St Bartholomew's and Trethowan at Guy's, and Rowley Bristow started one at St Thomas's soon afterwards.

At the beginning the work consisted of teaching on Tuesday morning in the wards, and on Wednesday afternoon in out-patients. The Dean directed students to attend these sessions, but not the operating session on Thursdays. There was never any lack of clinical material, for South London doctors had large panel practices in a densely populated area extending from Bromley to Vauxhall. In addition patients were sent to the orthopaedic department by old Charing Cross men, former residents from H.S.C. and from many doctors who had known him during the war. The accommodation in out-patients was adequate. For a short time Jennings Marshall was junior surgeon and early in 1922 was replaced by Buxton. The registrar was an out-patient officer and little else. He did some casualty work and helped in all surgical out-patient clinics. For a time Blackstock, who later did orthopaedics in Wolverhampton but died too soon, held this post. The house surgeon did other work, particularly as one of the casualty officers—a dual appointment of remarkable value. The out-patient teaching session was essentially for undergraduates, but usually others attended.

Even in the early days there were some unusual and most interesting patients. After the undergraduate teaching there was a short visit to the x-ray department and then a higher level discussion on the afternoon's work, which was greatly enjoyed by all the staff and visitors. Bellis Clayton, who was in charge of the Department of Physical Medicine and the Director of the School, was always there. It has appeared to me that important matters were discussed and the very close liaison between the two departments developed at these meetings and has persisted at King's, to the advantage of patients and students not only of the medical but also the physiotherapy and occupational therapy schools. Students of the ancillary departments as well as trained staff have attended in- and out-patients as a routine from early days.

In out-patient teaching Fairbank required a concise history, record of routine examination and notes of the radiographs, generally made by himself, irrespective of any report by a radiologist. The polite remarks to parents were always masterly; students enjoyed his methods of dealing with refractory children, and nurses the courtesy they received. This description indicates a standard set in routine but there was little delay in innovations.

A fracture clinic was started. All fractures diagnosed in casualty were radiographed and reduced and splinted by the casualty officer. On the following Tuesday all were seen at the fracture clinic, conducted alternately by the two orthopaedic surgeons, who taught the students, at first attending voluntarily, and informed the casualty officers, who were present, of their work. It may be of interest to note that plaster splintage was not used in the early 'twenties for Colles's and many leg fractures. Fairbank's teaching regarding ankle fractures was explicit and when he advised operation he did this with great skill and good end-results. However, the greater number of patients with leg fractures were treated as out-patients. For many years those with the major fractures were admitted under the surgeon on duty. However, the surgeon on duty often asked Fairbank to take over the case, or the junior orthopaedic surgeon treated many from the outset. In fact, by 1930, the fracture service at King's was well known in South London and patients were usually in casualty within an hour or two of the injury and seen shortly after by a member of the staff or a competent assistant.

The out-patient and fracture departments are today working on the same lines initiated by Fairbank, but there has been a many fold increase in the number of patients, accommodation and staff.

All who watched or assisted him operating know the high standard of technique he employed, based on the teaching of Lane. Although the " no touch " was adhered to, the gloves were always changed if there was any question of contact with unsterilised or possibly infected material. Probably he was at his best in the treatment of congenital dislocation of the hip, and in routine operation on the knee. He was meticulous in his operative technique for making an " upper lip " in children with C.D.H. Many came to King's around 1930 to see him do this operation. He was anxious in the cases when drainage tubes were used after operation for bone infection. When in usual good form in the theatre, the sister was addressed as Madam or Ma'am! Patients on whom he had operated were invariably visited within forty-eight hours.

His greatest help to King's was the organisation of a department, the inception and development of a fracture clinic and the collaboration of his department with that of physical medicine. There was no narrowness in this planning, and expansion was easy as the basic plans were sound. Comparable progress was made elsewhere and the pattern is in existence at many clinics.

Queen Alexandra's Hospital, Millbank—The Director-General of the Army Medical Services after the first world war realised that the consulting surgeons and Regular Army staff at Millbank required the help of specialists. Fairbank did years of work there and periodically went farther afield, especially to the Cambridge Hospital, Aldershot. Many officers have told me of the help he gave and how he would go at short notice at the end of a long day. Although his work was greatly valued, there was no conception at a high level of an orthopaedic service should an outbreak of war occur. Fairbank's work was clinical and it appears that his opinion was not otherwise sought.

" Treloar's "—Owing to the foresight, planning and generosity of Lord Mayor Treloar this hospital for children was opened in 1904. Henry Gauvain (later in life to be Sir Henry) was the first medical superintendent. The history of the successful hospital and training college is as well known as that of Oswestry, Biddulph Grange and The Heritage, Chailey. Sir Thomas joined the committee in 1924 and worked there for over twenty years. His first operating list was in April 1925, before he retired from Great Ormond Street, and he last operated there in September 1946. He helped the staff of Alton at his regular visits, but he had an interesting

appointment in being consulting orthopaedic surgeon to Portsmouth Orthopaedic Clinic and the Hampshire County Council. This entailed the care of in-patients (children only for many years) at Treloar's. In the course of this work he helped to train a number of young surgeons working there, including W. H. Langston. V. H. Ellis (St Mary's) joined him in 1946, but unfortunately his premature death robbed Alton of valuable services.

84 Harley Street—After nearly five years' absence from London his consulting practice had disappeared, but there was only a short period before it returned and increased rapidly. Fairbank followed the pattern of the time in keeping patients' notes in leather-bound books. As an assistant I was privileged to refer to these at times and have always regarded his notes as models for orthopaedic records. These notes were always up to date and there was a succinct report on the radiograph. Not infrequently there was a cryptic remark to his own credit or discredit, or on the doctor or possibly patient. It is probable that a feature of his private work was that other surgeons, general surgeons and orthopaedic surgeons were often at "84" with their patients, knowing that help was more likely there than in any other consulting room. It was a happy and elegant room with a large desk and differed somehow from most rooms—possibly in its simplicity.

But "84" was not only a consulting room. There were many opportunities to talk "shop." There were meetings there to discuss orthopaedic projects and " special clubs." Lady Fairbank and he entertained so many and in such a charming manner that surely anyone who had lunch or dinner there would remember the pleasant and cordial welcome and hospitality in rooms which they planned and allowed others to enjoy as much as they did themselves.

Fairbank has always had the fortune to remember people and places and he taught himself to make notes—generally on slips of paper—on the cogent matters to be dealt with in the daily round. However, he remembered many details of patients' notes and clinical findings. This applied particularly to the unusual skeletal abnormalities. An apparently new one would come before him and out came the remark, " Yes, that must be like the case I showed at the R.S.M. some years back. That one came from D. H., an old house surgeon at Ormond Street." The record was found with a reference to yet another one.

The reader will realise that assistance is required not to " jog this type of memory " but to help the machinery of recording so that all the workings ancillary to the master mind are left in a high standard of efficiency.

Miss B. Florence Bradbury—" Sister Bradbury " to many and " Sister " to others—has known Sir Thomas since 1907, when she went to the Hospital for Sick Children to train as a nurse, has been his third hand since 1919 and is still helping him to solve the problems coming to him. It is known to surgeons that during this century there have been a few women who have helped the development of surgery, perhaps particularly orthopaedic surgery, by being remarkable assistants or masterly organisers. Many have been trained nurses or had some association with our profession but not had medical school training. It is fair to her and her chief to make suitable reference here and to give credit to her. She saw Fairbank work in the operating theatre at Great Ormond Street in 1908–09, when she was theatre probationer, and later in Dresden ward. Operations for congenital dislocation of the hip were done, as Lorenz had been in London, and he used Murphy's button successfully for intestinal anastomosis. She saw more of his work when Sister of Helena for two years before the first world war. In 1919 she started her career at " 84." She was his theatre sister for private work, preparing the theatre as he wished it and scrubbing up at almost all the nursing homes he used. She made plaster splints. She was secretary—telephone, letters, showing patients in and out of the consulting room—all occupations allowing little time to spare. In the 'twenties and 'thirties letters took up some time, for Fairbank was a painstaking writer and dictated at length and as there were no medical secretaries at hospitals, consultants took the letters to doctors home for the secretary to type. When he was out the typing had to be done, and early on the collection and storage of radiographs and hospital patients' notes commenced in

addition to the work of private patients. This demanded skill and memory and increased each year. Frequent reference for demonstration of patients at meetings, or for articles in journals refreshed the memory, but the value of this collection really became evident in lectures during the last fifteen years or so, and in the publication of the Atlas. Sister Bradbury took infinite trouble to know what was stored and where. Others will recollect, perhaps as well as I do, the discussion around 6 o'clock in the consulting room on something unusual. The bell would bring in Sister and the answer to the question put to her was so often immediate and informative. She trained herself wonderfully to the off-stage work and helped on the stage when asked. She was able to help in the war years of the Emergency Medical Service and with the final sorting before Sir Thomas left " 84." Her friends and acquaintances are glad she is still at work.

BRITAIN 1939–1945

At the time when Neville Chamberlain was experiencing his first travels by air—and those to Central Europe—Fairbank was an equally busy man. Living at Harley Street, he was engaged in a large consulting practice and doing regular work at Treloar's Hospital and Millbank. He was visiting King's and Great Ormond Street on the request of his friends requiring help, usually with less common skeletal diseases. From many parts of the country he was being requested to study reports and radiographs of such conditions not well recognised. Evenings were spent collating information on these and writing. Holidays were medical meetings and short periods fishing, usually badly chosen, when " there was no water," and week-ends at " The Mill." This lovely house in a bend of the river must have given him, his family and their visitors great relaxation and was the envy of many.

Unfortunately the life of that period for everyone was changed almost overnight, for few were able to appreciate that the smouldering fire was leading to a holocaust, and that everyone within our shores was to become a pawn in the second world war. Fairbank had served his country in South Africa as a civilian doctor with surgical leanings, as a Field Ambulance officer and consulting surgeon in the first world war. In 1939 he accepted an appointment under the Ministry of Health as Consultant Adviser in Orthopaedic Surgery. The Emergency Medical Service was planned during 1938 and in due course organised as a national health hospital service for civilian illness, for the treatment of civilian victims of war and for the greater part of in-patient care of the sick and wounded of the Services.

The evacuation from London and other cities took place at some speed so that the Ministry of Health hurried the reorganisation of the hospital services. Fairbank's work became national. There were appointed in 1940 regional advisers, such as R. C. Elmslie, Harry Platt and T. P. McMurray.

Harry Platt was particularly helpful to Sir Thomas because of the appointments he held and his knowledge of the North, and became Assistant Consultant Adviser in 1940. Fairbank was Chairman of the Advisers (Regional Orthopaedic) Committee which began to have regular meetings at the Ministry in 1940. As the war progressed the work of the committee developed and the chairman was assisted by others. After my return from the Middle East and before the invasion of the occupied countries, those attending the committee included the Service and Ministry of Pensions Orthopaedic Consultants, when they were not regional also (such as Robert Milne and Rex Diveley, representing the Royal Navy and U.S. Army respectively), the Consulting Surgeon to the Canadian A.M.S., Robert Stanton Woods (Physical Medicine) and Rhaiadr Jones (Rehabilitation). As chairman, Fairbank was responsible to the Ministry for the " national orthopaedic set-up " for conveying to the representatives of the Minister of Health the recommendations of his colleagues and for the negotiations arising from these. The committee members were active and an orthopaedic service of importance developed. Fairbank was not content to do this work from an office. For a long time he was, in addition, a regional adviser and not only visited this area but spent much time on tour in the country seeing, at first hand, the problems of each regional adviser. It will

be remembered that as the population was less segregated than in pre-war days the diagnosis and treatment of the ordinary fractures—apart from injuries connected with war—presented a problem, which was solved by classifying the service available for fractures into Centre, Fracture A and B Hospitals. Fairbank took great trouble about this. The chairman and this committee, in due course, were able to see large orthopaedic centres and satellite units, which did little more than first-aid and were constantly visited by experienced surgeons. Those who were called upon to organise the orthopaedic arrangements for the National Health Service were materially assisted by the work of Fairbank and his committee.

During the war years he reported in medical journals his anxiety concerning delayed union of fractures, especially when skeletal traction was used, and infection around foreign bodies, such as transfixation pins for this purpose. He was not satisfied with the technique employed for the operative treatment of fractures and reminded readers of the " no touch " technique.

At the Mill, 1937.

THE POST-WAR PERIOD

After many years of hard work and of national service in three wars many expected Fairbank would retire from active work. In point of fact these years were fully occupied with important work. He returned to consulting practice in Harley Street and did a considerable quantity of hospital work, especially at Treloar's (Alton).

He was in constant demand to put a label on the unusual case of skeletal disease which often necessitated country journeys. In so doing he added to his collection of notes and radiographs. Periodic reports with accurate notes of a little-described bone condition were published and in 1951 the Atlas was well received and acclaimed a masterpiece. His presence

at medical society meetings and functions was seen less often—not from lack of desire to attend—but all regretted and felt great sympathy for him in that physical disability made it impossible for him to exchange scientific knowledge with so many who appreciated how much he was missed.

In due course the sorting out of papers was completed and " 84 " closed down. Forthwith Lady Fairbank and he took ship to see the world and a married daughter and her family in a distant Commonwealth. A holiday, yes, but he was at work; not only in New Zealand but also in Africa he was welcomed by orthopaedic surgeons.

It is a happy thought that the post still brings him radiographs and letters asking for help and an equally happy one that he got out a rod in '55 and went to old haunts north of the Border.

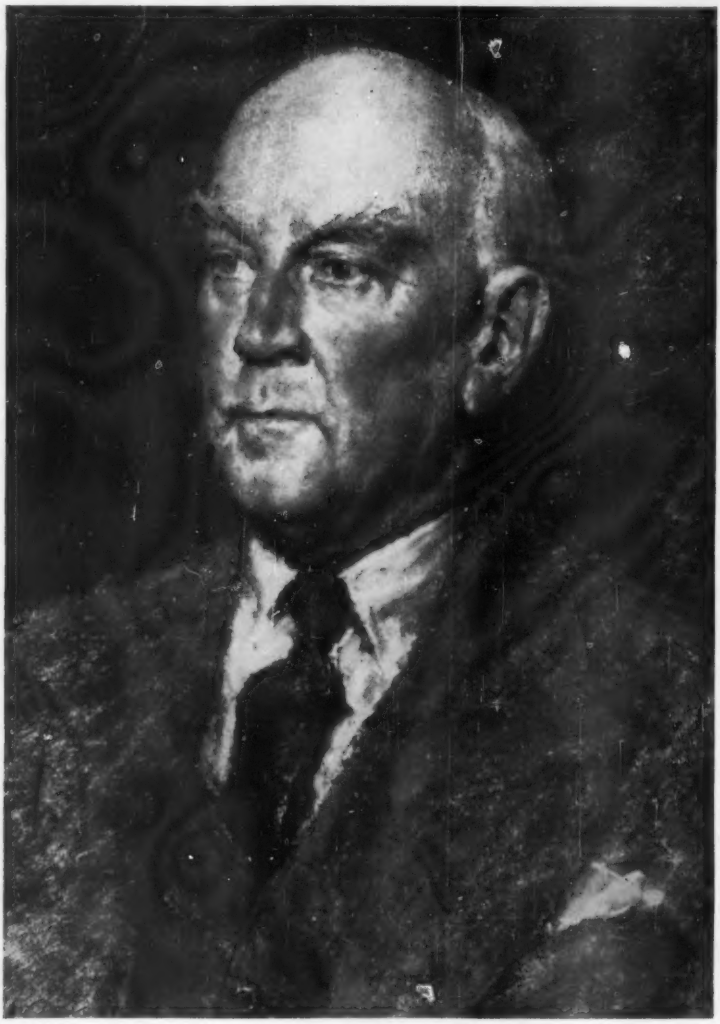
MEDICAL SOCIETIES

He has been a well known figure at the Royal Society of Medicine for nearly forty years—in the library and at meetings. He was elected an Honorary Fellow in 1951, and is a Past President of two sections. In 1922 the teachers of orthopaedics in London were active after the first world war in making a success of the Section of Orthopaedics. Fairbank, Elmslie, Bristow, Trethowan, Aitken, Bankart and Laming Evans set an example by regular attendance, bringing members of their teams. The latter were encouraged to show the cases under the care of the chief. An attendance of twenty or so grew rapidly, until the Section was one of the most popular at Wimpole Street, and on occasions in the late 'thirties the Hall was overfull. Fairbank brought to these meetings his experience, his friendship to the younger men, and a bag of notes and x-rays. He gave a number of addresses, always well illustrated, took part in many discussions, and in so doing was one who set the standard. Before and after these meetings he invariably had a word or two with the younger men, who always received encouragement at the meetings of this Section. A policy had been established that the meetings would form an excellent training ground for the younger men to demonstrate and discuss cases and read papers. In the early days most of the patients sent up had undiagnosed conditions. As knowledge of orthopaedics increased, surgeons sent groups of cases with end-results of treatment. Fairbank enjoyed both, and so often demonstrated some child with fantastic abnormality of bone and he always managed to show radiographs of every bone that mattered. Further I doubt if he showed any which were below a good standard.

His Presidential Address to the Orthopaedic Section was on " Some affections of the epiphyses " (1924) at a time when attempts were being made to classify " Schlatter's knee," Kohler's disease and other epiphysial abnormalities. Subsequently, as President of the Section for the Study of Disease in Children, he spoke on " Some remarks on osteogenesis imperfecta " (1930). In this paper he gave reasons for regarding the ante- and post-natal cases as due to the same disease.

The British Orthopaedic Association was constituted in February 1918. Fairbank was unable to attend the meeting as he was on military service with the British Salonika Force. He was elected to the Nomination Committee for 1918 and 1919, when there were twenty-one members. He was vice-president during much of the five-year presidency of Robert Jones and was himself president 1926–27. He is now an Emeritus Fellow, three of whom are founder members, Harry Platt and Arthur Rocyn Jones being the other two. The subject of his Presidential Address given on October 22, 1926, was " Some general diseases of the skeleton " (1927). He made a classification of general affections of the skeleton and of general disorders of the skeleton due to endocrine disturbance. At that time he stated " I cannot as yet discard the infective theory " as far as Paget's disease is concerned, but he had received a letter from Robert Lovett (just before his death) telling him that there were four monkeys in the Philadelphia Zoo, which had developed Paget's disease, one of them having been fed on a vitamin-free diet. The address will be discussed later.

Fairbank was a most regular attendant of the meetings. He enjoyed talking to all who brought problems to him and was always anxious to be a host. He was popular at the Continental meetings and had firm friends in many of the capital cities visited. He was more interested in the clinical demonstrations, operating sessions and papers and the chat with members than in the deliberations regarding the planning, teaching and development of a



A pastel by James A. Grant, R.P., 1952.

British pattern of orthopaedic surgery, which was discussed by the officers of the Association and were of the greatest importance in the 1920s. As the Association enlarged, English-speaking visitors and a number from Europe, notably from France and Italy, came to see Robert Jones and other surgeons in the centres. As has been mentioned, the Association was visiting overseas. The opportunity for the formation of an international society was becoming imminent. Efforts originating in 1913 culminated in 1929 in the foundation of

the International Society of Orthopaedic Surgery, now called SICOT (Société Internationale de Chirurgie Orthopédique et de Traumatologie). Fairbank was greatly interested and was a founder member, one of the three members of the first British Committee, and held office as vice-president of the second Congress in Bologna and Rome.

LECTURES

In 1929 Sir Thomas delivered the Lady Jones Lecture at the University of Liverpool on " Congenital dislocation of the hip, with special reference to the anatomy " (1930). Reference will be made to this and other papers and lectures, but here is quoted his closure of the address: " Is it too much to hope that the time will come, and that before many years, when every uncomplicated congenital hip will be cured at an early age, and the difficult problems presented by the irreducible case, hopelessly crippled on reaching adult life, will cease to worry the surgeon? "

Nearly ten years later he gave the first Robert Jones Memorial Lecture at the Royal College of Surgeons. The subject, " Increased and decreased density of bone, with special reference to fibrosis of marrow " (1939), was attracting the attention of surgeons, radiologists and pathologists. The diseases producing the conditions were classified and a table compiled of the calcium, phosphorus and phosphatase values in the blood in the various diseases. He drew attention to the frequency of Paget's disease, and the exaggeration of the number of patients who subsequently are affected by sarcomatous change. There is reference to the " Darmous animal," but at that time this type of osteopetrosis was not classified as fluorosis (Atlas, p. 316).

The Alex. Simpson-Smith Lecture is a memorial to a surgeon who lost his life near Tobruk after Rommel captured the garrison including 13 General Hospital, where he was surgical divisional officer. He was on the surgical staff of the Hospital for Sick Children, where this lecture is given.

Fairbank delivered this lecture on " Recent knowledge of affections of the skeleton in children " (1952) twenty-four years after he retired from the active staff. He made frequent reference to cases brought to him or reported by physicians and surgeons of the hospital. The remarks on infantile cortical hyperostosis and Engelmann's disease are of particular interest.

His subject was "Some unusual causes of backache" (1937) for the Founder's Lecture of the Chartered Society of Massage and Medical Gymnastics (now of physiotherapy).

CONTRIBUTIONS TO LITERATURE

In considering Sir Thomas Fairbank's contribution to surgical literature, one is impressed by his writings on congenital dislocation of the hip and on diseases of the skeleton, particularly those about which little was known thirty years ago. He has contributed many papers to the journals published in this country, of which a number may appear academic, but all had an impact on clinical thought. Review of them at this time reveals the pattern of his thoughts, the constant desire to report progress and follow up the fate of changes recognised and treated in the child. Throughout there is a high standard of literary expression, the subject matter being accompanied by illustrations well chosen.

At the end of this essay will be found a list of the articles and other contributions to literature. It is of interest to group certain of these and study such a group, for this brings to light the value of Fairbank's work. Many of the articles contain original Tables, usually in connection with differential diagnosis or end-results of treatment; the majority include clinical details of unusual cases.

Congenital dislocation of the hip—Information on diagnosis particularly is given in two papers (1928, 1929). The former was published and subsequent to a lecture entitled " The diagnosis of a limp," given to the Norfolk and Norwich Medico-Chirurgical Society. It is remarkably comprehensive, and the few hundred words on congenital dislocation of the hip ideal postgraduate teaching. *Inter alia* it is pointed out that certain signs are not diagnostic, but that the " diagnosis is usually quite simple if only the examination is made in a routine manner." The age when this is applicable is not stated. Then he proceeds: " Trendelenburg's sign is not diagnostic of congenital hips.

" The patient, stripped, stands with her back to the examiner and is told to lift first one foot and then the other from the ground. The position and movements of the gluteal folds are watched. When she stands on the normal leg, raising the other off the ground, the gluteal fold on this side rises slightly with the limb in the normal manner. When, however, she stands on the affected limb the gluteal fold on the sound side falls instead of rising. This is because the glutei on the affected side are unable properly to support the pelvis. This indicates nothing more than a defect in the osseo-muscular mechanism between the pelvis and the femur . . ."

The object of the second paper was to call attention to the fact that congenital dislocation can be diagnosed before walking is begun. Putti had shown (1929, 1933) the results of educating practitioners and mothers in early diagnosis.

The morbid anatomy was described in the Lady Jones Lecture and subsequently published (1930). This is a comprehensive paper of considerable value, which should be read periodically by surgeons dealing with this condition. The anatomy of the dislocated hip and of the muscles and the pathological changes are considered and illustrated by reproductions of radiographs, line drawings and paintings of specimens in the Musée Dupuytren. The latter are unusual and very realistic. It is possible they are a more permanent record than the author or artist realised at the time, as the curator of the Musée Dupuytren a few years ago was only able to find three of the specimens.

The last part of the article enables the reader to understand the views on treatment in 1930. It is stated that the objects of treatment are: 1) to provide a stable joint and obliterate the limp; and 2) to diminish the chances of arthritis supervening. His own work and others for the young child is reviewed and there are critical remarks on the shelf operation. Finally consideration is given to the irreducible case, in which increasing disability and pain develop in adolescent or adult life, and arthrodesis, osteotomy, the shelf operation and excision of the femoral head are discussed.

Shortly after the first world war Fairbank was anxious to publish the results of the treatment of congenital dislocation of the hip at the Hospital for Sick Children up to 1916. This was not possible at the time of publication of " Late results of treatment of congenital dislocations of the hip " (1922). In this are excellent paragraphs on each recognised form of treatment and the complications. The six conclusions—he states " they seem justified "—appear to be as true today as thirty years ago. When he spoke at the Royal Society of Medicine on treatment (1939) it was natural that to routine treatment he added " Simple abduction, increased gradually till extreme, and then maintained for a few months " (Putti 1929). With many years of experience he summarised the indications for open reduction or exploration of the joint and for the shelf operation.

The knee joint—Articles were written on the knee joint, particularly in the 1930s. Operations on this joint had become frequent, and little-described conditions were being found to be more common than had been realised. Osteochondritis dissecans was reported at the Centenary Meeting of the British Medical Association and the paper was published in 1933, but he stated the name was not justified by the etiology. The condition was described in several joints and cases affecting both knees that had been under the care of Rowley Bristow and of Somner were mentioned. The etiology was discussed at length and operative treatment advised. Eric Lloyd was the joint author in a report on cysts of the lateral cartilage of the knee with erosion of the head of the tibia (1934). By that time cysts of cartilages were

recognised and it was known they were more frequent on the lateral side of the knee, but that they could be responsible for damage to other structures by pressure was little known. The large experience of Sir Thomas enabled him to call attention to the chief differences to be found in a series of cases of derangement of the knee joint in young patients compared with those of adult age, in a paper entitled " Internal derangement of the knee in children and adults " (1937). There is an interesting account of eight cases of slipping patella and of a greater number of congenitally abnormal lateral cartilages. At the time of the latter there had been under his care thirteen below and three over the age of twenty. The illustrations in this paper are reproductions of his own line drawings and exemplify six types of abnormal cartilage.

General diseases of the skeleton—Early in his surgical career Fairbank was fascinated by the variety of general skeletal diseases. In the early part of the century the recognition and classification of these was of little interest to physician or surgeon and radiography was in its infancy. At the Hospital for Sick Children there appears to have been a collaboration of the staff to investigate the causation, diagnosis and treatment of these. In Fairbank's writings there is frequent mention of consultations with physicians and radiologists on a patient falling into this group of affections. By the autumn of 1926 Sir Thomas was able to give a presidential address of " Some general diseases of the skeleton " (1927), when he said he was referring to a few of the problems. He realised that many of the doubtful questions would not be solved for years to come and that the clinician must be joined by the biochemist and pathologist. The work of Robinson, Kay and Soames had been published; so a beginning had been made in the value of calcium and phosphorus in metabolism. Fairbank stressed the importance of metabolic and endocrine errors and of the tests employed to establish a defect.

Teamwork was necessary to make progress, and Fairbank collected the evidence, sifted and studied it and asked, time and again, for more evidence, even if it became necessary to wait until another patient with comparable signs was brought to his notice. Probable additional radiographs and Miss Bradbury's searches for other case records were important in the factor required for differential diagnosis. As time went along Fairbank came to be recognised as the authority in this country not only on the unusual local bone condition—particularly at the end of a bone and in a child—but on general skeletal abnormalities, that at one time might have been under the care of a paediatrician. The flow of patients and case records increased and the retirement from active work at hospital appears to have been associated with an increase of this. Papers continued to interest paediatricians, surgeons, radiologists and a few pathologists, not only in this country, and an international reputation was well deserved. Most fortunately Sir Thomas set to work after the second world war to co-ordinate his writings and add to them. The result is well known and greatly appreciated, for *An Atlas of General Affections of the Skeleton* (1951) is a record of a great life's work. Surely it is well known in the clinics of all English-speaking nations. Certainly many conditions reported are of little more than academic interest, but knowledge of them makes the life of the scientific surgeon of such a great interest. Until this publication the young man seeking information on rare skeletal conditions did not know where to look. Now he is sure to find enough information and excellent illustrations of every affection, whether it be Paget's disease, Fanconi's syndrome or Cooley's anaemia. The case reports are fascinating, and there is never too much on one subject. Some conditions were first described by Fairbank. He took trouble to find a suitable name, but there is a clarity of the condition by his choice of name. He has made accessible information that was little known and it is beyond our reach to feel envy at his great accomplishment.

Many will join me in saying " Happy birthday and thank you for much advice."

I wish to express my thanks to many friends for information that I have included in this essay.

A CHRONOLOGICAL LIST OF SIR THOMAS FAIRBANK'S CONTRIBUTIONS TO SURGICAL LITERATURE

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CORNER, E. M., and FAIRBANK, H. A. T. (1904): Sarcomata of the Alimentary Canal: with the Report of a Case. Practitioner, **72,** 810.

and in (1905): Transactions of the Pathological Society of London, **56,** 20–42.

(1905): Gall-Stones and Cancer. (Correspondence.) Lancet, **i,** 1,542.

(1905): A Discussion on Toothache, Neuralgia and Remote Affections of Dental Origin. British Medical Journal, **ii,** 559.

(1905): Cases of Sprengel's Shoulder. Transactions of the Clinical Society of London, **38,** 200.

(1905): The Treatment of Necrosis of the Jaws. British Journal of Dental Science, **48,** 1,059–73.

(1905): The Treatment of Congenital Talipes. Clinical Journal, **27,** 139–44.

CLOGG, H. S., and FAIRBANK, H. A. T. (1905): Appendicitis at Charing Cross Hospital. Statistical Tables of Cases of Operation for Appendicitis during the Years 1902, 1903, and 1904. Medico-Chirurgical Transactions, **88,** 517–24.

(1907): Two Cases of Injury of the Head in Children. Lancet, **i,** 89–90.

(1907): Sarcomata of the Jaws. British Medical Journal, **ii,** 1,506–8.

GARROD, A. E., and FAIRBANK, H. A. T. (1907): A Case of Catarrhal Appendicitis due to the Presence of the Oxyuris Vermicularis. Lancet, **ii,** 772.

(1907): articles in An Index of Treatment by Various Writers. Edited by R. Hutchison and H. S. Collier. Bristol: John Wright and Co.: also later editions.

(1908): A Discussion on the Surgical Treatment of Infantile Paralysis. British Medical Journal, **ii,** 909.

(1908): A Clinical Lecture on Congenital Dislocation of the Hip and its Treatment. Lancet, **i,** 1,752–60.

(1909): A Case of Coxa Valga. Transactions of the Medical Society of London, **32,** 352.

(1910): A Clinical Lecture on Lymphadenitis. Clinical Journal, **37,** 170–6.

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(1911): A Case of Bilateral Congenital Dislocation of Patella after Operation. Transactions of the Medical Society of London, **34,** 402–3.

(1911): A Case of Endothelioma of the Gum. Proceedings of the Royal Society of Medicine (Odontological Section), **4,** 12–13. *And in* British Journal of Dental Science, **54,** 110–11.

(1911): Cyst of the Jaw, the Result of Traumatism. Proceedings of the Royal Society of Medicine (Odontological Section), **4,** 14–15, 17.

(1912): A Clinical Lecture on Appendicitis in Children. Medical Press and Circular, **N.S. 93,** 664–6.

(1912): A Demonstration of Surgical Cases at the Polyclinic. Clinical Journal, **40,** 106–12.

Includes the following:

Osteo-myelitis; Diminished Growth of Tibia; Talipes Varus, 107–8.

Obstetric Paralysis; Operation on the Brachial Plexus; Subspinous Subluxation of the Shoulder-joint, 109–10.

Obstetric Paralysis; Birth Palsy, 110–11.

Infantile Paralysis of the Shoulder, 111.

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FAIRBANK AS A SOLDIER

JULIAN TAYLOR, LONDON, ENGLAND

It was in 1907 that the writer first met this outstanding surgeon and from that year until 1918 it was his good fortune to be under Fairbank's command or within hail of his advice. This was not just a matter of military authority or of Fairbank's high place in surgery, though both were powerful forces: it was that he found himself always with Fairbank to look up to. In this he was not alone for it was the general attitude of all juniors and many seniors.

In this year 1907 Fairbank, already a volunteer soldier of some seniority for he had served in the war in South Africa, was Transport Officer of the London Companies of the R.A.M.C. Volunteers, and in this horse transport section the writer, with other very junior medical students, enlisted as a driver. Those were the days of horsed ambulances and general service wagons such as had been used in South Africa and with these and other contemporary medical equipment we trained at Aldershot, Richmond Park, and elsewhere. Naturally we did not meet our officer on terms of military equality but we rapidly learned that he was kindness itself to his soldiers and we saw that he was a good horseman with the lightest of hands. His obvious affection for our horses was clearly returned by them during the fortnight's holiday from their normal post-office duties that our annual training afforded them at Aldershot or on Salisbury Plain.

It was not until the summer of 1914 that the writer received his commission in this unit, having risen from the rank of lance-corporal, but immediately after qualification he approached Fairbank asking to become his clinical assistant at the Hospital for Sick Children. The reply was that he usually had Fellows of the College as his assistants, but his sense of obligation to his soldiers may have made him waive his rule in favour of the writer who had in fact passed his final examinations a few days earlier. If this be so, it is probably the only occasion in his life on which Fairbank put the interests of anyone before those of his patients.

At Great Ormond Street it is true we were subjected to a different kind of authority but it was exercised in the way that the writer already knew, there was no sensible difference. Nevertheless Fairbank's out-patient clinics at Great Ormond Street were quite unforgettable, for they combined an already world wide authority (in particular on the subject of congenital dislocation of the hip) with the same kind and sane approach to mothers, children, infants, and doctors, that we recognised in his dealings with non-commissioned soldiers and horses. It is probably unnecessary to say that it often needed Fairbank to discern the virtues of parents and children, or if he was too busy to do so it was his habit to assume their existence.

BELGIUM AND FRANCE

Then came the war of 1914 by which time the writer had become an officer and perhaps a few lines should be written about the unit to which Fairbank gave so much and in which he served with distinction before and during the early half of the war. With the formation of the Territorial Army, the R.A.M.C.V. London Companies had become the 3rd London Field Ambulance of the 1st London Division. The volunteer soldiers who made up the unit were of many types and from all classes but it is fair to say that nearly every member, young or old, was actuated by the same sense of duty in joining. Among them were a few medical students, though the exigencies of the war soon deprived us of them. It is probable that this unit was the first Field Ambulance that ever existed as a formation, those of the regular army not existing in peace but being formed on mobilisation. The officers were partly London consultants and among surgeons there were the late Major W. E. Miles who

left us for other duties before the war started and the late Major F. B. Waggett, two very different personalities each in his way pre-eminent. There were also Captain Fairbank and Lieutenant R. M. Vick. Lieutenant Bernard Potter was anaesthetist to Charing Cross Hospital which was also Waggett's and Fairbank's, though Waggett was originally from Cambridge and St Bartholomew's Hospital. In addition there were two junior officers lately from the House at St Bartholomew's Hospital, the late Lieutenant F. H. Robbins and Lieutenant (now Major-General) R. E. Barnsley. Thus four members of the mess were Bart.'s men and it was soon apparent that, if one must go to war, Bart.'s men make good companions. This is recalled here and these names are mentioned because the writer is sure that Fairbank was also conscious of a piece of good fortune.

The mess was a very friendly one with " Uncle Tom Fairbank " as an older member of the younger group. All of us looked up from the beginning to this man who seemed to us to be the essence of wisdom, kindness, commonsense, and unselfishness. Like most units of 1914 we were anxious to be off to the war but our prospects did not seem to be rosy. The god from the machine turned out to be Fairbank who it seemed was a god with seven machines for in some way he collected the funds for the purchase of seven motor ambulances, vehicles which were new to the establishment and of which the army was extremely short. How this unpublished patriotic gift to the war effort was organised has not been told, but our suspicion is probably correct that Mrs Fairbank (as Lady Fairbank then was) and her family had practical hands in it. When we were about to leave for Belgium they were delivered to us and we then learned that not only had Fairbank organised their purchase but had also had the forethought to order them with an extra 2 ft. 6 in. in the chassis so that there should be no overhang behind the back axle. It is probable that these were the best and most comfortable though not the most showy motor ambulances of that war. The gift of motor ambulances did our business for us and we became the 85th Field Ambulance in the 28th Division. Fairbank had two functions for he commanded a section of the unit and was also Transport Officer; it was not until much later in the war that we were sent an Army Service Corps officer to take over transport duties. Our clearest memory of those days is the very unpleasant winter of 1914–15 in Belgium where our division took over an uncomfortable and all too warlike length of line in front of Ypres. We were a division of regular soldiers, who made up for their ignorance of trench-warfare by their many admirable qualities in adversity and success. In the ditch of knee deep half-freezing mud that was their line they were everywhere overlooked by the enemy who, still sore at his failure to break through in October, were perpetually active in the offensive. The heavy casualties by wounds and trench feet were disasters which reduced the division seriously and one of the writer's most vivid memories of the time is that of an officer with frozen trench feet and a shattered arm. His wounds were being dressed by Fairbank, dressings being supposed to be the sole surgical duty of field ambulance medical officers at that time. As soon as he felt Fairbank's gentle touch on his wounded limb, he breathed contentedly " At last the expert." Perhaps he never met another just so gentle and so expert.

The second Battle of Ypres burst upon us with the first gas attack, and then dragged on, with failure to achieve the enemy object but with much bloodshed and unhappiness on our side. All over our division and outside it, Fairbank's position was now consolidated as the great surgeon with the Field Ambulance who also looked after the horses. With the end of the second Battle of Ypres veterans remember that the salient had become greatly reduced in area and the problems of transport thereby considerably increased, because there was not now a corner that the enemy could not see. At the same time the B.E.F. began to reorganise itself to some extent. The difficulties were not lessened when the wisdom of the doctrine of early evacuation of the wounded to safer areas began to be questioned, the institution of forward operating units being contemplated.

About this time it was discovered that there was no orthopaedic surgeon at the base in

France but a large number of soldiers knew that there was one serving in a forward area in Belgium and that he was devoting his attention to techniques of transport rather than to surgical treatment. Accordingly it was decreed that Fairbank should be transferred as orthopaedic specialist to a general hospital then landing in France. However our A.D.M.S. was loth to lose a good officer and the transfer did not take place. Already in 1915 an order was becoming a basis for discussion rather than a command. What may be the truth behind the countermanded transfer is probably known to no one other than its subject, who was and is the soul of unselfishness and discretion, but it is likely that had the discussion been carried a little higher, nothing could have stopped the transfer. The probable simple truth is that Fairbank was a good transport officer, fond of horses, happy in his loyalty to his Field Ambulance and soldiers, and did not press for change.

GREECE

The rest of the unit's history in France and Flanders during 1915 is that of active trench warfare combined with preparation for, and the eventual failure of, the Battle of Loos, with the now familiar mud, blood, cold, and misery. After watching for a month the ferocity of the battle subsiding into the waste of angry necrosis that we had come to regard as inevitable, we were sent to another part of the war in Salonika. So we handed over our prized ambulances (without overhang) and on arrival in Greece received in exchange some model T Fords (with a great deal), together with some light cavalry horsed ambulances and about a hundred mules. These latter beasts, unpromising as they seemed in their untrained state, surprisingly soon under Fairbank's supervision, learned to carry the pack-saddles and cacolets, the litters and travois, to which we were to entrust our wounded. There followed a pause in our active warfare and we spent a very cold and uncomfortable winter in Greece.

With spring, which is beautiful in those parts, came green grass and with it the announcement of the divisional horse show, which turned out to be a good replica of such festivities at home. There had been much sickness among the troops, partly due to the poor quality of their food, one result of German submarine activities in the Mediterranean. Accordingly during the whole winter and spring our ambulance horses had been overworked and were thin and scraggy, while in contrast those of the gunners were fat with shining coats for they had had little work to do. The appearances of our beasts earned us a severe reproof from H.Q.; indeed the only beast of Fairbank's section to receive approval was a blue roan mule that was still so bad tempered that we could find no use for it. It was so unsocial that there was only one driver that it would permit to come near it, kicking and biting everyone else. It was shown by him and commended for its fine condition, due no doubt to plenty of food and no work.

Following this we suddenly departed to the Struma Valley for the Bulgars had appeared at the Rupel Pass. To the Struma our brigade had to make a road to supplement the " carriage road to Seres " described by Baedeker. The new road was necessarily constructed at express speed by troop labour. At one point a battery had difficulty in getting a gun across a rocky obstruction and the whole column behind was held up. More and more teams were tried but the fat and shining horses all jibbed. A polite offer of help from Fairbank was accepted somewhat to our surprise and four of our condemned screws easily pulled the 18-pounder over the difficulty. The Divisional C.R.A. did not permit this brigade of Field Artillery to forget the incident.

It was about this time that Fairbank gave up his responsibility as transport officer, so here a characteristic and revealing story may perhaps be permitted. During the cold winter of 1915–16, while at his morning toilet, he saw an old Greek woman leading along the road a very lame and overloaded donkey, with one leg in a splint. Uncle Tom walked over, looked at the donkey and commanded that a picketing peg be brought. The picketed donkey

was then unloaded by the Transport Sergeant-Major, the usual dreadfully sore back was exposed, and at the same time the cause of the lameness was seen to be a fracture. Uncle then without a word produced his revolver and shot the donkey while the old Greek woman in a frenzy chattered curses and imprecations. Uncle, with a few sympathetic words that she did not understand, then pressed upon her a bundle of hundred drachma notes, far more than the donkey was worth, and then went on with his shaving.

Our next troubles were those connected with the disastrous outbreak of malaria which appeared punctually in June and for which the Struma Valley had been notorious since the time of the Romans. To Fairbank fell the duty of transporting thousands of sick soldiers, mostly suffering from subtertian malaria, many miles to the Seres road where they were picked up by lorries and taken 70 kilometres to Salonika. This he did most efficiently, pitifully though his own soldiers were reduced by sickness and death and though the choice was not easy of the sick fit to make the long road and lorry journey.

A little later in the year there was an outbreak of active fighting in the Struma and it occurred to authority that Fairbank was a surgeon, so after the battle was over he was transferred to the 29th General Hospital as surgeon. By the end of the following year he had become assistant consulting surgeon with the late Colonel R. E. Kelly as his senior.

Shortly before this the writer had also been transferred to surgical duties in a C.C.S. and had begun to work under Fairbank's direction. The change from anti-malarial sanitation to surgery was sudden, and it was years since he had had a surgical thought. He is permanently indebted to Fairbank, whose good sense, thoughtfulness, surgical judgment, ever tendered help, and endless other qualities, guided him continually during this period when he was learning the clearing station surgery, and he remembers very clearly how the consulting surgeon could always be summoned from Salonika, fifty miles away.

Fairbank's appointment as Consulting Surgeon to the Salonika Army occurred at a time when there was continuous fighting near Lake Doiran. It also coincided with a change in the surgical treatment of wounds for it was now established that the serious and often fatal infections of the wounds inflicted in the early battles could be avoided by prompt excision. Policy was tending in the direction of excision followed by primary or delayed suture of the large majority of wounds. It was the writer's privilege to be among the surgeons who carried out the change of policy under Fairbank's direction. It was of inestimable value to have the consulting surgeon's opinion always available at short notice. It also gave a satisfactory feeling of confidence to be able to rely on Fairbank's outspoken support should military authority become critical of the amount of forward surgery being done by relatively junior officers. He was indeed an ideal consulting surgeon to serve.

The consulting surgeon overworked himself keeping a constant check on the wounded from clearing stations to base hospitals. He also invited his forward surgeons regularly to the base to see for themselves what happened to their patients and to hear his comments. This kind of ever watchful sanity was the special quality of his work. Informed as it was by his great surgical and orthopaedic knowledge, experience and judgment, there was nothing in it nor in him that had any quality of the showman. His was a personal service to the sick and wounded conducted by willing deputies. The necessary result was that while he did not spare himself, the work did not spare him either.

The final attack on the Bulgars which ended the war was for the British a frustrating and particularly bloody one during which Fairbank again showed his qualities in the service of the wounded and of those who were tending them. He contracted malaria and his final efforts wrecked his health for the time being, but by now the task was ended and the war soon over.

Many people have heard that Fairbank was in charge of mules instead of surgery in the 1914–18 war. It is the writer's hope that the preceding lines have suggested that although this was true for two years, his time was in no sense wasted. For a man of his parts, of his

invariable good sense and, let it be said, of his outstanding straightness, must by his qualities influence his neighbours. In the bad business of war when opportunities for lower standards of behaviour present themselves daily, such an example is as valuable as it is exceptional. Others will no doubt write in worthy tributes of his eminence as an orthopaedic surgeon. How widely and deeply his personal influence was felt in the 28th Division and later among the surgeons of Salonika, how, in short, he was respected by everyone from general to private, is the index of the value of his presence and in the writer's opinion no higher tribute could be paid. He feels sure that inadequately though this greeting may be expressed, survivors of the 28th Division, of the 85th Field Ambulance, and of his medical colleagues in the 1914–18 war, will join in expressing their heartfelt thanks for what he was and for what he did in that campaign. On his eightieth birthday we wish him many happy returns of the day.

A REVIEW OF ORTHOPAEDIC SURGERY IN BRITAIN

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In the year that Queen Victoria ascended the throne a young physician in England introduced cutting for the correction of deformity. Tenotomy of the Achilles tendon was in itself a simple procedure but it was a new departure in the cure of crippling; its success was sufficient to warrant W. J. Little's embarking upon a close study of the etiology, pathology and rational treatment of deformity. To achieve this purpose hospital accommodation was essential; he found, however, that general hospitals were too preoccupied with a multitude of other ailments to give time or space for the problem of crippling in the community. He therefore aroused the interest of the public to the deplorable state of the crippled poor, and the response was such that in 1840 the Orthopaedic Infirmary was opened in Bloomsbury. In casting about for a name descriptive of the particular function of the institution Little adopted the word orthopaedia (" straight child ") coined by Andry in 1741 from two Greek words. In this way the word orthopaedic was added to our English vocabulary—a term expanded in its meaning by the process of time. It is worth recording that British usage in writing the word has remained faithful to Greek etymology and has refused to pay homage to a confusing phonetic expediency. The work of the Infirmary grew so rapidly that by 1845 it moved to Hanover Square, to a building with greater accommodation, and was granted a Royal Charter of Incorporation wherein its name was changed to the Royal Orthopaedic Hospital.

Orthopaedic surgery in England literally began on the threshold of the Victorian Era. The first decades were occupied in an intense study of the etiology and pathology of deformity as a scientific basis for treatment. Little and his disciple William Adams were particularly active in their investigations, each having a distinctive approach of his own to the problem of crippling. Little, cured by Stromeyer of Hanover of a paralytic equinovarus foot due to poliomyelitis, felt compelled to seek facilities for the treatment of cripples in England. But he remained a physician, although keenly interested in the investigation of deformity and in such correction as was obtainable by splinting, manipulation and tenotomy. His book, entitled *A Treatise on Club Foot and the Nature of Analogous Distortions*, published in 1839, is a classic. His lectures on deformities of the human frame were collected and published exactly a century ago and his famous paper on infantile spastic paraplegia was an important contribution to neurology and orthopaedics.

The study of deformities was commended to Adams by his great teacher J. H. Green, the philosopher surgeon of St Thomas's Hospital, who encouraged him " to take up the new study of orthopaedy because it opened a wide field for pathological investigation and usefulness." And when elected surgeon to the Royal Orthopaedic Hospital in 1851 Adams was already a well disciplined pathologist of considerable experience. With great energy he explored the whole field of deformity and wrote extensively upon it. What he wrote bears the impress of first-hand knowledge and research. He published a monograph entitled *On the Reparative Process in Human Tendons*, based on research on rabbits and observations of tenotomy in human beings over a period of nine years. He spent several years investigating club foot, its causes, pathology and treatment—work which secured for him a Jacksonian Prize. He made an exhaustive study of spinal curvature and published a book upon it; he asserted that " horizontal rotation of the bodies of the vertebrae always precedes lateral curvature." His comfortably fitting spinal support has never been surpassed. Adams made wide use of subcutaneous tenotomy, employing it with great success in the treatment of wryneck and Dupuytren's contracture.

For many years correction of deformity depended entirely upon stretching, subcutaneous cutting of soft tissues, and splinting, but eventually distorted bone was also attacked. Subcutaneous osteotomy was introduced by Langenbeck in 1852 when he corrected a flexion ankylosis of the hip by blindly cutting the neck of the femur. It is interesting to note that the first surgeon to adventure on osseous correction in England was Stromeyer Little (so named in honour of the surgeon who treated his father), who in 1868 at the London Hospital operated upon a patient aged fourteen years whose both knees had a fixed right-angular flexion, the left due to fibrous ankylosis and the right due to osseous fusion. The left knee was straightened by gradual stretching on an extension apparatus and the right by subcutaneous osteotomy. A carpenter's chisel was inserted through an incision a third of an inch long over the lateral femoral condyle and driven by mallet across the bone. The chisel was several times withdrawn from the femur, but not out of the wound, and each time driven through intact bone until the femur could be broken manually. The hamstrings were severed and the limb was nursed on an extension apparatus. " The result of the treatment related above was satisfactory as the girl could walk easily without assistance."

Although subcutaneous osteotomy had been available for some of the more severe deformities yet its uses were limited. Open operations were forbidden. Hence the whole practice of surgery was very restricted and its progress retarded. There was no lack of dexterity in the art of operating, but post-operative sepsis, pyaemia and hospital gangrene caused such heavy mortality that little beyond emergency surgery was thinkable. Almost up to the last quarter of the nineteenth century there was no security, even for the simplest of operations, against any of these catastrophes. A surgeon having closed his wound was, in the parabolical words of Richard von Volkmann, " like a husbandman who, having sown his field, waits with resignation for what the harvest may bring, and reaps it, fully conscious of his own impotence against the elemental powers, which may pour down on him rain, hurricane and hailstorm." How small was the field of surgery is revealed when it is recalled that in 1865 there were 397 surgical beds at St Bartholomew's Hospital and yet the average annual number of operations during the preceding five years was only 370, of which seventy-eight were amputations.

ANTISEPTIC SURGERY

In 1867 Joseph Lister announced in the *Lancet*, and at the Annual Meeting of the British Medical Association in Dublin, his Antiseptic Principle in the practice of surgery. It was the most astonishing and beneficent gift to humanity since the dawn of medicine. It was so revolutionary and contrary to the age-long conception of healing that many of the surgeons of Lister's generation were unconvinced of its value or frankly repudiated it. But Lister continued steadfastly experimenting in his laboratory for the improvement of the application of the antiseptic system, at the same time endeavouring to persuade the profession to adopt it; the main reason for his leaving Edinburgh for King's College Hospital was the opportunity afforded of demonstrating the great virtue of antiseptic surgery to the London schools. The younger surgeons, free from prejudice, became converted and it was they who really embraced the new system and started the phenomenal advance in surgery. In the forefront was Lister's dresser, William Macewen of Glasgow. He perfected linear osteotomy by antiseptic surgery and devised for the purpose a special bone-cutting instrument, bevelled on both sides, which he called an osteotome. In 1879 he succeeded in promoting the reconstruction of a humerus, most of which was destroyed by osteomyelitis, by sewing along the bed of the absent shaft bone chips removed from other patients for the correction of deformity. This was the first time that bone had been successfully transplanted from one human to another—an achievement of fundamental importance for orthopaedic surgery and one which stirred the imagination of T. H. Huxley when Macewen communicated a paper to the Royal Society upon this work. Furthermore it started Macewen on thirty years of clinical and experimental investigation on *The Growth of Bone*, culminating in 1912 in his great book of that title. Another pupil of Lister,

Thomas Annandale of Edinburgh, explored the knee joint for internal derangement and, finding a torn cartilage, sewed it back in position which resulted in comfortable restoration of function of the joint. For a surgeon to have been able to explore the knee joint without any inflammatory reaction or any delay in union of his wound was a good test of the efficacy of antiseptic surgery. During the years immediately following Herbert Allingham of St George's Hospital evolved a complete technique for exploration of the knee joint with removal of loose cartilage when present.

The dozen years from 1870 onwards were a peculiarly flowering period in orthopaedic surgery, for apart from the initial impact of antisepsis and its effect on bone and joint surgery there were other contributions of importance. Sir James Paget gave an exhaustive and complete description of a deforming inflammatory disease of bone which he called osteitis deformans, and he drew attention to the occurrence of a non-inflammatory avascular necrosis of bone as the result of trauma. In 1875 Hugh Owen Thomas published his classical monograph on *Diseases of the Hip, Knee and Ankle Joints*. It at once revealed him as an original thinker in surgery. The burden of his teaching was rest for diseased joints, conveyed in his often repeated formula " enforced, uninterrupted and prolonged rest." For Owen Thomas rest had a precise meaning. It could only be obtained by striving for immobility. This he endeavoured to achieve by his ingenious but simple splints. In the period that he wrote he undoubtedly secured by rest and fresh air such healing of tuberculous joints that hardly occurred outside his own practice. His famous caliper, originally designed for joint disease, was subsequently used for fractures of the lower limb. Nothing is more symbolic of the genius of Owen Thomas than this splint, surprisingly simple in construction but peculiarly adaptable in its uses. It is a basic equipment wherever fractures are treated. The modern use of the caliper is frequently combined with traction by weight and pulley, an indispensable aid to skeletal surgery introduced for the first time by John Haddy James of Exeter and described by him, with illustrations of its use, in an address on surgery delivered at Liverpool in 1839.

Pasteur with his revolutionary germ theory of disease had prepared the way for the introduction of Lister's antiseptic system. And Koch, stimulated to seek what maladies were of microbic origin, found in 1882 that a bacillus was the cause of scrofula and phthisis, thereby proving their unity. The importance of that discovery was beyond compute in its significance for human well-being. It provided a scientific basis for all the various forms of treatment that followed and for the study of the domestic and social environment of those affected by tuberculosis. And although the tubercle bacillus has proved peculiarly resistant to attack its immunity shows some sign of weakening when confronted by some of the later antibiotics. At long last this protean disease is clearly on the decline in Britain.

OPEN OPERATIONS ON BONE

When orthopaedic surgeons had grasped the practical details of antiseptic surgery it was natural that they should begin cautiously with its application in osteotomy, where a small skin incision was adequate. It was not until the 'nineties that bolder surgery began. Robert Jones started performing arthrodesis of the ankle and knee for severe infantile paralysis, and by 1894 he was able to report fifteen patients upon whom this operation, introduced by Albert of Vienna in 1875, had been carried out. He spoke more favourably of this operation for the ankle than for the knee. In 1908 he described his well known two-stage arthrodesis operation for paralytic talipes calcaneo-cavus. Again Muirhead Little advocated cuneiform tarsectomy for resistant talipes in adults and adolescents, after a successful removal of a wedge from the neck of the talus and calcaneum. But a leading orthopaedic surgeon, with some alarm lest others should follow Little, felt obliged to issue a warning that " only those who are perfectly sure of their antiseptic methods should undertake tarsectomy."

The other procedure to arouse attention was tendon transplantation for infantile paralysis, introduced by Nicoladoni in 1881 when he attached the peroneus longus and brevis tendons

to the tendo calcaneus for paralytic calcaneo-valgus. The patient, when shown six months later at the Salzburg Surgical Congress, exhibited well restored plantar flexion of the foot. But it was not until 1897 that tendon transplantation was first performed in England, when Frederick Eve at the London Hospital operated upon four patients with paralytic feet, one with talipes equinovarus in whom the tibialis posterior was attached to the extensor digitorum longus and a strip of the tendo calcaneus was attached to the peroneus longus; in two months the child walked well. The following year T. H. Openshaw wrote an excellent paper on tendon transplantation, full of detail indicating the selection of tendons for transplanting, rules for successful grafting, the cause of failure, and the best suture material with comment on its preparation. In 1903 appeared the classic monograph *Surgery of Paralysis* by Tubby and Robert Jones. The application of the new surgery to the whole field of flaccid and spastic paralysis was well covered. Tendon transplantation and arthrodesis were considered separately and in combination. About the most successful of all transplantations is that for radial nerve paralysis evolved by Robert Jones during the first world war. Congenital and paralytic deformities claimed most of the interest of orthopaedic surgeons; the potential crippling of trauma had not yet attracted their particular attention. Fractures were usually treated in the casualty departments of hospitals; patients with severe injuries were admitted as in-patients. But the standard of treatment, judged by the average functional result of a limb fracture, lagged behind that obtaining in other branches of surgery. For many years a fractured femur, whether of neck or shaft, was treated by tying a long Liston splint to the body and lower limb with possibly strapping extension. Fibrous union of a fractured femoral neck, with its attendant instability, was an inevitable expectation. There were a few surgeons, however, who were disturbed by this inadequacy of surgery and who regarded it as their duty to restore to an individual the full use of a limb broken by accident. Prominent among them were Hugh Owen Thomas and Robert Jones, to whom flowed the casualties of Merseyside and those incident to the cutting of the Manchester Ship Canal. Stress was laid on the accurate re-apposition of broken ends of bones, and on the restoration of true alignment, by powerful traction and skilled manipulation under anaesthesia. The limb was subsequently immobilised in a caliper or in an arm splint modelled on the caliper, with accessory malleable gutter splints or collar-and-cuff.

After the experience of a decade in the handling of fractures Arbuthnot Lane had reached the conclusion that for the intractable fracture open reduction with rigid fixation was necessary. Speaking of simple oblique fractures of the tibia and fibula in certain classes of labouring men he stated: " I have no hesitation whatever in asserting that, under the methods of treatment at present adopted, not only is the man totally incapacitated from earning a living for an unnecessarily long period but in a considerable proportion of cases he is unable subsequently to perform such heavy work as he was able to do before the injury, so that he is obliged to follow some less remunerative pursuit if indeed he has not to depend solely on charity." On January 8, 1894, Lane operated upon a man with oblique fractures of tibia and fibula after failing to reduce them by manipulation under anaesthesia and subsequent splinting for a few weeks with no sign of healing. " The fracture in the tibia was exposed by an incision 4½ inches long when its direction was seen to be very oblique running downwards, outwards and backwards. Several small fragments of bone and muscle intervened between the ends of the bones . . . After much difficulty the broken surfaces were brought into accurate apposition by means of lion forceps and two screws were inserted." Thereafter he persisted with the operative treatment of fractures. In an ununited fracture of the neck of the femur he obtained osseous union by passing two screws from the greater trochanter through the neck into the femoral head, thereby anticipating the later nailing operation. It was sometime after 1905 that he introduced his well known steel plates and screws; with these he extended the range of fracture operations.

Lister in 1877, with antiseptic precautions, wired a fractured patella, obtaining osseous

union, a result rarely obtained by external splinting; but despite the success of the operation he was subjected to some unfavourable criticism for this " unjustifiable procedure." And Lane by his operating on simple fractures raised a storm of criticism and even abuse. He soon realised that if the operative treatment of fractures was to be safe rigorous asepsis was essential; he therefore evolved an unremitting no-touch technique which of itself has had a transforming effect on operative orthopaedic surgery.

There is one of the lesser disabilities, which is very common and often very painful, that is deserving of notice—namely hallux valgus. For this there was no radical cure until 1885. In that year J. N. C. Davies Colley of Guy's Hospital performed his operation of removing the proximal part of the first phalanx, whereby the great toe was straightened, the patient relieved of symptoms, and the treading power of the head of the first metatarsal preserved. Various modifications have been adopted since, including the removal of the head of the metatarsal, but for the severe type of the deformity Davies Colley's operation, with the removal of the prominent inner part of the metatarsal head, has stood the test of seventy years.

Many of the advances in medicine have been dependent upon discoveries of pure scientific origin. In 1800 Sir Humphry Davy produced complete experimental evidence of the power of nitrous oxide to induce safe anaesthesia. Pasteur, another chemist, proved the microbic origin of disease in plants and animals; and in December 1895 W. K. Röntgen, a physicist of Würzburg, announced the discovery of x-rays which, among their many uses, have enabled surgeons to obtain a knowledge of the pathology of living bone and joint afflicted by disease or injury and to gain precision in some orthopaedic procedures. A voluntary worker in Liverpool, on receiving the *Frankfurter Zeitung* containing an article on Röntgen's discovery, translated the narrative for Robert Jones, who at once crossed to the continent and brought back an x-ray apparatus by means of which he and Thurstan Holland took a picture of a boy's injured wrist. It displayed a small embedded bullet; the two marvelling at the revelation.

The advances in orthopaedic surgery led to a greater interchange of ideas among younger surgeons. At the annual meeting of the British Medical Association in Bristol in 1894 an informal meeting was held with the object of founding an orthopaedic society, and on November 3 of the same year the British Orthopaedic Society was formed, the avowed object of which was the advancement of orthopaedic surgery. Forty members were enrolled. Three meetings a year were held. The first took place on January 31, 1895, at the Royal Medico-Chirurgical Society's rooms. Subsequent meetings were held at Liverpool, Birmingham and Bath, among other places. But in spite of its stimulating discussions the Society only lasted a few years; four slender volumes of its transactions were published. It had served a useful purpose because it had brought together men who were interested in orthopaedic surgery, and the memory of this coordinated effort lingered. They had first met together at the rooms of the Royal Medico-Chirurgical Society and they were destined to meet again fourteen years later in the new house of the same Society, renamed the Royal Society of Medicine, to form an Orthopaedic Sub-section of the Section of Surgery.

EMERGENCE OF ORTHOPAEDIC SURGERY AS A DISTINCT SPECIALITY

At the beginning of the twentieth century it was assumed that a surgeon who had completed the customary apprenticeship was able to deal adequately with almost any surgical condition for which there was a known remedy. This attitude was encouraged by the romantic experience of the 'nineties during which the full impact of anaesthesia and antisepsis on surgery was felt; it was a decade of adventure. Most teaching hospitals felt that there was no need for special departments, although two or three of them delegated a surgeon, as a part-time appointment, to the duty of taking charge of ophthalmic or orthopaedic patients. But Jonathan Hutchinson, with clear foresight, declared that selective specialisation would become inevitable; the expanding knowledge of surgery, with its multiform technique, would be beyond the power of one man to encompass. The first indication of the advancing status of orthopaedic surgery,

in a hospital service, occurred at Charing Cross Hospital in 1906 when Mr H. A. T. Fairbank was appointed orthopaedic surgeon in succession to F. C. Wallis, who had been promoted surgeon. Sir Frederick Wallis had been appointed assistant surgeon in 1893 and in the following year was also given charge of an orthopaedic department. His main interest, however, was in rectal surgery, in which he achieved great distinction and was the author of a manual on that subject. When he became full surgeon he resigned his post as orthopaedic surgeon. Mr Fairbank was surgical registrar, and the Board of Governors were anxious to retain his services permanently for the hospital, but there was no vacancy on the ordinary surgical staff; they thereupon appointed him, on March 14, 1906, to the vacant post of orthopaedic surgeon. He differed from his predecessor in that he had no surgical duties outside his own department of orthopaedic surgery. It was an unparalleled appointment. Six years later similar appointments were made at St Bartholomew's and at Guy's Hospitals.

At that time there were scarcely a dozen surgeons in England who maintained a constant interest in skeletal disabilities, but the following half-century has seen a remarkable change. During this period Sir Thomas Fairbank has been a leading influence in moulding the shape of orthopaedic surgery in Britain; indeed he has come to hold a unique place in the heart and mind of all those who follow the same calling. His vitality and sustained interest in orthopaedic surgery are a delight to many. He has played an honoured part in the service of his country, in hospital organisation and as an advisory servant of the State. But his most personal and abiding influence has been imparted to his writings; in these he has always striven to base advancing knowledge on scientific fact derived from pathology. He has devoted most attention to skeletal disabilities such as congenital dislocation of the hip, coxa vara, and in particular diseases of bone which culminated in his greatest work, *An Atlas of General Affections of the Skeleton*, published in 1951.

His description of the anatomy of congenital dislocation of the hip is about the best on this subject that has appeared. He drew attention to the changes in the bone behind the acetabulum, with the occasional formation of a facet at this spot, to the arrangement of the capsule, and to the importance of the ischio-capsular band which forms a sling over the neck of the femur. The work is illustrated by many beautiful drawings of specimens examined by Sir Thomas in the Musée Dupuytren in Paris. The original drawings are now available among the archives of the British Orthopaedic Association. His long experience covers the whole period of treatment by manipulation in this country, for he was present at the demonstration by Lorenz at the City Orthopaedic Hospital in 1903. He has said that if the dislocation is reduced before the age of three years, or better still before the age of two, a permanent cure ensues in a very large percentage of patients. The hip will be approximately normal and stand the wear and tear of life almost, though not quite, as well as a hip that has never been abnormal. During the progress of treatment he laid great stress on one guiding sign, namely " The position of the femoral head with regard to the centre of the acetabulum is of greater prognostic value than the shape of the upper margin of the socket." Furthermore, even with the help of open reduction the age of six years for a bilateral dislocation and nine for a unilateral should not be exceeded. For the shelf operation the ideal patient was one of four or five years with a unilateral dislocation and unusually poor acetabular margins.

In his study of coxa vara he succeeded, by a process of close reasoning and the examination of many radiographs, in separating a particular variety of the congenital form of this malady to which the name of infantile or cervical coxa vara had been given. He pointed out the recurring characteristic triangular fragment of bone occupying the lower part of the femoral neck close to the head and which later became fused to neck and shaft. This pathological phenomenon he attributed to developmental error, a supposition that was supported by its presence in children with other irregularities of bone growth.

The " Atlas " is the rich harvest gleaned during a lifetime of patient observation, careful clinical recording and the collecting of innumerable radiographs. In this book the author

has gathered together from scientific journals his many contributions on those affections of the skeleton where more than one bone is involved. The introductory essay on these osseous maladies is a most valuable feature of the work, clarifying by intimate knowledge many vague notions about their pathology. A classification is drawn up based on their etiology, and tables are given which are helpful in diagnosis, particularly the one on blood chemistry. The Atlas is of supreme worth as a work of reference in a field of surgery in which uncertainty is apt to reign. It has a value beyond that of a period; it is one of the best scientific contributions that have emerged in British orthopaedic surgery.

In the years immediately preceding the first world war there were clear signs of an increasing interest and activity in orthopaedic surgery. In 1912 Macewen's classic *The Growth of Bone* and Tubby's great two-volume text-book appeared; in the same year St Bartholomew's and Guy's Hospitals formed orthopaedic departments independent of general surgery, with R. C. Elmslie and W. H. Trethowan respectively in charge. Elmslie was about the last to hold a resident appointment at the old Royal Orthopaedic Hospital in Hanover Square. He returned to St Bartholomew's to become chief assistant in an orthopaedic department which had been started in 1867, with an assistant surgeon usually in charge. He also spent five years in the pathology department gaining an experience that influenced his subsequent approach to all orthopaedic problems.

His work on the pathology of coxa vara, communicated in his Erasmus Wilson lecture of 1907 and in a subsequent monograph, is outstanding in its scientific presentation; it is the original source of much of our knowledge of this disability. He classified coxa vara into three main groups, those due to 1) injury, 2) bone softening from disease, and 3) idiopathic, which he subdivided into congenital, infantile or cervical, adolescent. All these varieties had already been recognised, but the infantile or cervical group was so named by Elmslie himself; in it he described the V-shaped gap in the upper part of the neck but was inclined to believe that this type of coxa vara was due to injury. Several years later Fairbank with good reason regarded it as of congenital origin. In describing adolescent coxa vara Elmslie marshalled convincing evidence to prove that it was due to trauma, stating that " instead of the original opinion that accidents to the femoral neck in children were rare and were due to severe injury, we must accept it as demonstrated that such accidents are common, that a very slight injury may be sufficient to produce them, and the immediate disability may be correspondingly slight . . . I have recorded a case of a boy in whom this accident occurred during a hurdle race, which he was able to finish and win, and cases in which the epiphysis is apparently separated by very slight wrenches of the hip joint, the deformity only showing itself some months afterwards, are comparatively common. The actual site of the injury in the neck of the femur has been a matter of some dispute. I have no doubt that during adolescence the injury is situated close up to the epiphyseal line of the head, but not of course actually through the epiphyseal cartilage, so that, technically speaking, the injury should be classed as a separation of the epiphysis."

Elmslie also made a special study of fibrocystic disease of bones, upon which he wrote a great deal. He communicated an extensive paper to the British Journal of Surgery in 1914. He examined a large number of specimens in many of the pathological museums of London, giving their clinical history and histological sections. He concluded that in calling the malady a fibrocystic disease did not imply a knowledge of its total pathology, for it was uncertain whether it was one disease or several, and whether it was an inflammatory process, new growth, or simply an instance of disordered bone growth. He wrote: " It would rather appear that there is a general disturbance in the processes of growth or alteration in the bone. That in the course of this there is 1) a fibrous metaplasia of the marrow; 2) an excessive formation and activity of the giant cells; 3) an active absorption of bone, with coincident formation of new bone and cartilage. That in places the fibrous mataplasia may be most evident, leading to the formation of fibrous masses; in other places cysts may be produced, either by the action

of the giant cells or by the degeneration in the fibrous tissue; and that in still other places there may be abundant formation of bone—which may or may not be properly calcified—and of cartilage or fibro-cartilage; whilst, last of all, the giant cells may be produced in such numbers as to form a veritable myeloma. These changes appear to have a period of activity, and then to become stationary."

We owe to Elmslie an extension of our knowledge of the pathology of bone disease, a subject by which he was ever fascinated. His orderly judicious mind enabled him speedily to select what was relevant to the solution of any problem that confronted him. In an open discussion he would express his considered opinion in a clear unfaltering style in a voice of considerable charm; all the more remarkable because, in spite of appearance, he was never really in full health. He exercised a profound influence on his younger colleagues, always inculcating upon them the fundamental importance of pathology in relation to orthopaedic surgery, a subject to which he turned again in his presidential address to the British Orthopaedic Association.

ORTHOPAEDIC TEACHING

The greater recognition of orthopaedic surgery by teaching hospitals secured its regular tuition to undergraduates. But before this period sound teaching in this branch of surgery had been available in several text-books, the best of which was published by A. H. Tubby in 1896. This work was an authoritative presentation of orthopaedic surgery as understood in the closing years of the nineteenth century; the author cast his net widely in order to gather the theory and practice of surgeons in America and the Continent. His own researches on tendon repair were described at length. In 1912 a new edition appeared with the ominous title *Deformities including Diseases of the Bones and Joints*. Owing to the advance of orthopaedic surgery between the two editions, and the author's deliberate inclusion of skeletal diseases within its range, the whole work was rewritten with the various subjects arranged according to their etiology and pathology rather than on a regional classification. It was issued in two large volumes; it contained seventy plates and more than a thousand illustrations, of which four hundred were original. In the following words Tubby boldly declared that " Orthopaedic surgery is the surgery of the entire locomotor apparatus. In England, contrary to the custom abroad, works written on ' Orthopaedic Surgery,' although they have dealt with tuberculosis of the spine, have not included tuberculous and many other forms of diseases of the bones and joints elsewhere. And this, in spite of the obvious fact that in the practice of Orthopaedic Surgery a surgeon is constantly called upon to deal with such morbid conditions and their results. It has been my attempt to remedy this somewhat illogical state of affairs, and to treat the subject from its actual standpoint, which is the surgery of the entire locomotor apparatus, dealing not only with actual deformities, but also with those morbid processes which involve potential deformity. It is not claimed that the scope of the orthopaedic surgeon's work is thereby enlarged, since he undertakes already the treatment of malformations and deformities of the muscular and osseous systems, which demands a full acquaintance with the pathology of such conditions and with the essential therapeutic methods. A considerable portion of the work has therefore been devoted to a description of arthritic and osseous diseases." This was the best text-book on orthopaedic surgery in any language and has scarcely been equalled since; the clear descriptive power of the author was not the least of its features. It raised the prestige of British orthopaedic surgery.

Tubby devoted a good deal of attention to tendon transplantation, to which he was first attracted by the work of Lange and Vulpius during his studies at Halle and Leipzig. He devised an operation by which the pronator radii teres was converted into a supinator; sometimes he combined this procedure with transference of the flexor carpi ulnaris to the extensor carpi ulnaris and of the radial flexor to the radial extensors. *The Surgery of Paralysis* (1903) by Tubby and Robert Jones was on a theme in which both were intensely interested; each had

made a pioneering contribution on tendon transference. Furthermore each was eager for the advancement of orthopaedic surgery and this book was a sign of their happy cooperation.

The developing interest in this branch of surgery was further stimulated by the holding of the Seventeenth International Congress of Medicine in London in August 1913. Never before in the history of orthopaedic surgery in Britain did so many leading orthopaedic surgeons from Europe and America assemble together as they did to the Section of Orthopaedics over which Robert Jones presided. The papers and discussions were fully representative of the whole range of orthopaedic surgery. Among those who took part in discussion were Lucas-Championnière, Ridlon, Whitman, Lovett, Brackett, Baer, Putti, Spitzy, Vulpius, Dollinger and Rollier. A paper that attracted considerable attention was that by Fred H. Albee on " Original uses of the Bone Graft as a treatment for Ununited Fracture, Certain Deformities, and Pott's Disease." At the Royal National Orthopaedic Hospital the technique of bone grafting was demonstrated by Albee before a crowded audience. With his motor-driven twin saw an autogenous graft was cut from a tibia and implanted in the split spinous processes of the vertebrae of a patient with Pott's disease. The pervading cordiality that prevailed at the president's dinner to the members of the section evoked no whisper of the deep cleavage that would occur in twelve months.

THE FIRST WORLD WAR

In August 1914 the thunder of a world war crashed, suddenly causing a deluge of casualties. The Army Medical Service made hurried improvisation to meet this bewildering situation. Leading physicians and surgeons were appointed to various commands. Tubby went out with the British Mediterranean Force and was stationed at Alexandria with Sir Victor Horsley. He gave a graphic account of the Gallipoli Campaign and of the sufferings of the troops. He organised fracture and orthopaedic treatment in Egypt but was unable to get sanction for segregation of fracture patients or for special hospitals for their treatment. In Britain Robert Jones was attached to the Western Command and in his tour of its hospitals he found that " no provision of any sort was made for cases crippled and deformed, and early evacuation was both instruction and the routine. The result was that many men were discharged from the Army in a very large number of cases totally unfitted either for military or civilian life." To stop this state of things he succeeded in persuading the War Office in 1915 to reserve 400 beds at the Military Hospital at Alder Hey, to which wounded soldiers requiring orthopaedic treatment might be transferred from other hospitals. The same year he went to France and gave demonstrations at the casualty clearing stations in the use of the Thomas splints; these splints became a permanent part of war equipment. The caliper in particular saved thousands of limbs and helped in the comfortable transport of the wounded. He also wrote two small manuals which had a widespread influence on all fronts, the one *Injuries of Joints* and the other *Notes on Military Orthopaedics*.

In 1916 the newly built Hammersmith Infirmary at Shepherd's Bush was converted into a military orthopaedic hospital similar to that at Alder Hey, and was staffed by younger orthopaedic surgeons, some of whom had been trained at Liverpool. Military orthopaedic hospitals were established later in other parts of the country; these were staffed by surgeons drafted from Shepherd's Bush and by relays of surgeons from America in charge of Goldthwait. This chain of hospitals provided fracture and orthopaedic treatment, with rehabilitation for the wounded soldier in reasonable proximity to his own home. This was a great achievement and was only accomplished after much obstruction; it was a great triumph for the persuasive persistence and energy of Sir Robert Jones.

The establishing of military orthopaedic hospitals with the appointment of Sir Robert Jones as Inspector of Military Orthopaedics was a clear indication of the capacity of orthopaedic surgery to prevent crippling from skeletal injuries and of its ability to help in restoration of manpower for battlefield or factory. And when later the military orthopaedic surgeon took

off his uniform on his return to civilian life he did not divest himself of his skill but was able to apply it to the accidents of industry. With the restoration of peace the special military hospitals began to disappear, but soon there arose others in another form like those at Oxford, Stanmore, Harlow Wood and Exeter—country hospitals following the pattern of the pioneer hospital of Robert Jones and Agnes Hunt at Baschurch and Oswestry with its network of clinics in adjacent counties. All these hospitals attracted younger surgeons in whom the war had cultivated a taste for orthopaedic surgery and who were anxious to gain experience in the care of civilians with skeletal disorders.

The military orthopaedic hospitals had diffused greater knowledge of the practice of orthopaedic surgery than at any other time, and the response of the younger generation was reflected in the crowding of scientific meetings for its discussion. It was this inspiring atmosphere that encouraged Robert Jones, with the help of Robert Osgood of Boston, to form a British society on the model of the American Orthopaedic Association. The inaugural meeting of the new British Orthopaedic Association was held at Roehampton House on February 2, 1918, with Muirhead Little as president. Little was a man of rare culture, beloved by patients and colleagues, with an encyclopaedic knowledge of orthopaedic literature. He took a leading part in establishing Queen Mary's Hospital at Roehampton as an artificial limb-fitting centre, and he wrote a monograph on *Artificial Limbs and Amputation Stumps*, and an admirable history of the British Medical Association for its centenary meeting in London in 1932. His wife started a hospital school at the National Orthopaedic Hospital as early as 1892, and its success was such that the local education authority became responsible for it. It was not until the Fisher Education Act of 1918 that education and treatment of physically defective children of school age became compulsory. About twenty founding members of the new Association were enrolled, forming a group most of whom had worked with Robert Jones. Under his leadership they enlarged the boundaries of orthopaedic surgery in Britain.

BETWEEN THE WARS

During the post-war decades there was remarkable progress in orthopaedic surgery. Through the military orthopaedic hospitals which he had organised Robert Jones gave convincing proof of the value of segregation, continuity of treatment and unity of control in the successful treatment of skeletal injuries—three principles he had learnt long ago when responsible for the treatment of casualties incident to the cutting of the Manchester Ship Canal. Furthermore the particular experience of the men chosen for the staffing of these hospitals implied that the care of fractures and allied injuries was a part of orthopaedic surgery, which is the surgery of the entire locomotor system. The first ostensible linking of fractures with an orthopaedic department of a hospital was made by Sir Harry Platt when he established a fracture clinic at Ancoats Hospital, Manchester. This was followed with a similar clinic by Sir Reginald Watson-Jones at Liverpool Royal Infirmary.

No-one contributed more to our knowledge of the operative treatment of fractures than Hey Groves. He conducted numerous experimental observations on bone grafting in animals, the results of which he embodied in his Jacksonian Prize Essay of 1916. He employed every form of bone graft and adopted many forms of fixation. He concluded that the " most ideal graft is a piece of living bone used in its entire thickness . . . The success of a living graft depends very largely upon: *a*) the extent of its contact with living bone; *b*) the accuracy of its apposition; *c*) the firmness of its fixation." He also made extensive researches upon internal splinting in animals, using plates and intramedullary pegs of bone and metal. He applied all this experimental work on bone grafting and metal splinting in the treatment of human fractures. The combined work was ultimately published in his book, *On Modern Methods of Treating Fractures*, an enlarged second edition of which was published in 1921. During the period of these researches the non-electrolytic metals had not yet been introduced, but it is interesting to note that he proved the effectiveness of the intramedullary steel peg in

fractures of long bones. Again commenting on the frequent difficulty of controlling the upper short fragment in transverse subtrochanteric fractures of the femur, he bound the fragments together in alignment by an intramedullary bone peg six inches long introduced at the top of the greater trochanter. By this procedure he anticipated intramedullary nailing. Hey Groves was largely responsible for the foundation of the *British Journal of Surgery*. Its high standard from its inception owes much to his tireless devotion as first editor for nearly thirty years; by this work " he built for himself his best monument."

Among the many problems of skeletal trauma those involving the shoulder have commanded a good deal of study varying from the stiffness of traumatic periarthritis and injury of the rotator cuff to that of frank dislocation. The treatment of recurring dislocation left much to be desired owing to uncertainty of cure by the operative procedures adopted. This was due mainly to operations performed without reference to pathology, although Curling of the London Hospital had long since discovered a capsular detachment and humeral defect in such dislocations; but his disclosure was forgotten. Blundell Bankart in 1923 rightly stressed the importance of basing treatment on pathology and pointed out the detachment of the glenoid labrum, advocating its reattachment and repair in all patients, supporting his contention by an impressive high proportion of permanent cures. Putti and Platt independently devised an effective and less difficult operation; subscapularis was detached an inch from its insertion and the distal portion attached to periosteum and soft tissues in front of the neck of the scapula with the arm rotated inwards; the proximal portion was sutured to tissue at the lesser tuberosity of humerus. This procedure checked lateral rotation of the humerus, which was the movement producing dislocation. Bankart's operation has stood the test of over twenty years and is the best known of his contributions. He exerted great influence in the development of orthopaedic surgery in Britain. As surgical registrar at Guy's Hospital he assisted Arbuthnot Lane, learning from that master surgeon a rapid dexterity and a faultless no-touch technique. He was a quick operator, who neglected nothing, and was a most accomplished surgeon, for he combined neurosurgery and children's surgery with that of orthopaedic surgery. For several years he performed all the neurosurgery at Maida Vale Hospital for Nervous Diseases. He was about the first to perform Stoffel's operation for spastic paralysis, enabling very many children to walk and attend special schools, and one of the first in this country to perform autogenous bone grafting. He planned and rehearsed an attack on difficult territory with meticulous care; he cured by radical excision patients exhausted from chronic intractable sinus-riddled tuberculous hips. He was so absorbed in the art of operative orthopaedic surgery that he inspired his pupils to aim at excellence in its technique.

The modern teaching of skeletal injuries was gathered up in a comprehensive text-book by Sir Reginald Watson-Jones entitled *Fractures and other Bone and Joint Injuries* published in 1940. It was the best work on the subject that ever appeared and it was not without significance that it was written by an orthopaedic surgeon.

Although the war had focused attention on traumatic disabilities yet other orthopaedic problems soon attracted interest, amongst which were disorders of the foot. Paralytic instability occupied a prominent place. In 1922 Naughton Dunn introduced triple arthrodesis, including excision of the navicular bone, for calcaneo-cavus and other deformities, combining it when necessary with tendon transplantation. Dunn became lecturer in orthopaedic surgery at Birmingham University after a training at Liverpool and a period as surgeon at Shepherd's Bush. For paralytic calcaneo-cavus Elmslie performed a two-stage operation, the first stage corrected the cavus deformity by soft-tissue stripping and talo-navicular arthrodesis, the second corrected calcaneus by arthrodesis of the posterior part of subtalar joint, fixing a strip of the tendo calcaneus to the tibia and attaching the peronei, flexor digitorum longus and tibialis posterior to the back of the calcaneum. In 1927 Lambrinudi published his ingenious operation for drop foot, a procedure he evolved while directing a small clinic at

Balaam Street, East London, before his staff appointment to Guy's Hospital. The antero-inferior part of head of the talus was removed as well as the postero-inferior quarter of the navicular. The remaining part of the head of the talus was then insinuated into the gap in the navicular; the subtalar joint alone was arthrodesed. Lambrinudi had a penetrating intellect and expressed hinself in an original and stimulating fashion on many orthopaedic problems. He held that there were " no principles of orthopaedics as opposed to principles of medicine. There were only principles of medicine, which in orthopaedics should be applied to the restoration and maintenance of the function of locomotion, using the term in its widest sense. Always he laid stress on function as the only true guide in treatment and not only function of the body as a whole but also its proper functioning in relation to its social environment."

A foot deformity of recurring interest is congenital club foot, for which manipulation and splinting have been the basic means of correction. Denis Browne produced effective splints of simple design although they lacked the anatomical precision of the older and more expensive appliances of Scarpa and Adams. For the stubborn or neglected deformity, which could not be completely corrected by manipulation, open section of resisting or shortened ligaments, the so-called soft-tissue operation, was adopted by Elmslie and Trethowan and also by Brockman, who reinvestigated the pathology of club foot, emphasising that it was of the nature of a subluxation inwards of the navicular bone and interior part of the foot around a fixed muscle-free talus. Trethowan, because of his abhorrence of the wrench as an instrument of correction, was all the more disposed to lay bare the tarsal ligaments and cut or excise what he saw offending. Trethowan, an ardent disciple of Arbuthnot Lane, confessing that he himself was not naturally dexterous, by sheer disciplinary practice became the outstanding orthopaedic craftsman of his time. To see him bone grafting or openly correcting and fixing a difficult fracture of the ankle was an exhilarating experience. Like his master he was notorious for the length of his incisions but remarkable for the dryness of his wounds; he would say: " if you get down to the bone at its most superficial part and stay there you cannot cut anything of importance."

Until the beginning of the century congenital dislocation of the hip was considered incurable, but interest revived in this deformity after the demonstration of the Paci-Lorenz manipulation at the City Orthopaedic Hospital in 1903. Jackson Clarke took a leading part in fostering manipulative reduction and retention by plaster. He was highly successful in his late results and published a monograph upon the subject. After the war attention was directed to the dislocation that resisted manual reduction or in which the femoral head was unstable after reposition. For this type of hip Sir Thomas Fairbank and Sir Harry Platt advocated exploration to remove capsular obstruction with or without the prising down of an oblique ill-formed acetabular roof. In 1927 Putti, with his exceptional experience of this almost racial malady, successfully campaigned to get infant patients under the age of one year to the Rizzoli Institute where he was able to reduce a dislocation by gradual abduction on a divaricator. The success of this procedure influenced the surgeons at the Wingfield-Morris Orthopaedic Hospital to extend continuous traction to older infants, using an adjustable abduction frame for the purpose.

Reduction by gradual traction or manipulation, with retention by plaster or special splint, is still the method of choice in Britain. This procedure is justified by the high proportion of permanent cures achieved by surgeons in the earlier days of manipulation. Laming Evans in particular obtained results that were unsurpassed chiefly because of his skill as a manipulator and his meticulous after-care. A child with a unilateral dislocation was permitted early walking in a plaster with a high patten on the shoe of the affected side. When the plaster was discarded a $1\frac{1}{2}$-inch patten was worn on the shoe of the sound side to promote abduction of the opposite limb; up to two years the child slept in a removable plaster with the thigh abducted 90 degrees with some flexion. Again he would by patient repeated manipulation produce an astonishingly

good correction of a badly deformed club foot where a less disciplined junior would seek relief by open section. Evans had a sensitive easily provoked personality and was deeply conscious of his duty to his patients, who loved and feared him alternately but who continued to crowd his clinic. For the advancement of orthopaedic surgery he bequeathed £20,743, the ultimate residue of his estate, to the Royal College of Surgeons, whose Council from the income has instituted a research scholarship known as the Laming Evans Fellowship.

The first surgeon in Britain to study and treat tuberculosis of all joints on any considerable scale was Hugh Owen Thomas. Before he announced his famous hip and knee splints for joint disease he had tried his methods and checked his results on more than a thousand patients. He was fully alive to the value of rest, good feeding, fresh air and sunshine in the cure of joint tuberculosis. Even in dwellings of the very poor he insisted on nursing children out of doors on beds improvised for the purpose. Robert Jones continued the conservative tradition of Thomas when excision, flushing and curetting were common practice.

One of the earliest hospitals for children suffering from surgical tuberculosis was opened at Alton in 1908 and named after its founder the Lord Mayor Treloar Cripples' Hospital. Sir Henry Gauvain was appointed superintendent, and under his guidance the hospital achieved a national reputation. He arrived at Alton to find an advisory medical board still reluctant to give up radical operations, but Gauvain went to Berck-sur-Mer to study conservative open-air treatment and the technique of making and using plaster splints. He collaborated with Calvé in advocating aspiration instead of free incision for evacuation of tuberculous abscesses. He was convinced of the value of sunbathing, and was a pioneer in promoting heliotherapy as a means of increasing resistance to the disease, but insisted on graduated exposure under strict supervision. He established a hospital school and a college for occupational training. Gauvain's happy and optimistic temperament infected staff and patients.

Koch's discovery of an invading bacillus revealed tuberculosis to be one malady whatever its manifestation, and we now know that joint disease is very frequently secondary to primary disease in the chest. It has long been customary to segregate pulmonary patients in sanatoria, and with the development of thoracic surgery to aim at rendering a patient non-infective on his return home. The establishment of country orthopaedic hospitals provided greater facilities for the treatment of patients with skeletal disease, hospitals like those at Margate and Sevenoaks having done pioneer work. Greater bed accommodation for the study of joint tuberculosis led to progressive knowledge of its treatment. In 1924 George Perkins wrote his prize monograph on *The Diagnosis, Treatment and End Results of Tuberculous Disease of the Hip Joint*. This work was based on an experience at the Country Orthopaedic Hospital at Pyrford. It was an able disquisition into the clinical nature of the disease and of its treatment, introducing a new abduction frame for prolonged conservative treatment. It was found that destructive disease of the femoral head very often ended in fibrous ankylosis of the joint, leading to an unreliable weight-bearing limb; a hip with osseous ankylosis at 35 degrees flexion was the best functional end-result for most purposes. This was difficult to obtain, because primary intra-articular operation was not in favour with English surgeons. This ideal was attained some years later by an extra-articular bone graft bridge from greater trochanter to ilium. And in 1932 H. C. Trumble of Melbourne achieved ischio-femoral arthrodesis by a tibial graft. H. A. Brittain followed with a solid fusion obtained by a Trumble-McMurray procedure; a broad tibial graft was passed along the track of a displacement osteotomy of the femur to impinge in a prepared cavity in the ischium. The tendency to adduction in hip disease pressed the graft towards the pelvis; this arthrodesis is well adapted to bear the stress of weight bearing. Brittain fused other joints by so placing his bone grafts that they were subject to compression force during use of the limb. His early death was a loss to rational orthopaedic craftsmanship.

The commonest site for skeletal tuberculous disease is the spine; it is also the part that

well illustrates the inherent power of recovery. In earlier days it was not uncommon to see individuals with a kyphosis whose spinal radiograph disclosed a healed destructive disease which could have been no other than tuberculosis and for which no treatment was received. On the other hand it is the place where the most distressing paralytic complication may occur, particularly in the mid-thoracic region, where the normal kyphosis is greatest and where the calibre of the spinal canal is smaller than elsewhere, thus allowing little accommodation for débris or exudate. Spinal tuberculosis with paraplegia was first accurately described by Percivall Pott in his monograph of 1779. He differentiated between flaccid and spastic paralysis and noted that spasticity was the invariable rule of spinal cord pressure in vertebral caries. He concluded that the disease had its origin elsewhere in the body and was capable of revealing itself in a variety of organs—or, in modern terminology, tuberculosis is an infective disease with local manifestations. Continued recumbency in a plaster bed brings relief to most patients but there is a minority for whom operation is necessary. The earlier attempt to remove spinal cord pressure by laminectomy was discouraging because of its uncertain relief and its high mortality. As a substitute for laminectomy Ménard in 1894 introduced costo-transversectomy. Transverse process and rib of diseased vertebra are removed, thus permitting a gloved finger to be cautiously insinuated between rhythmically moving pleura and vertebra until the abscess is reached; fluid pus and débris are evacuated without opening the spinal canal. For the following forty years Ménard's operation was favoured as the best radical procedure in the event of failure of paralytic recovery by recumbency. But further interest in paraplegia of Pott's disease was greatly stimulated by R. W. Butler and H. J. Seddon in their intensive studies of its etiology, pathology and treatment. About this period, in 1933, a patient with Pott's paraplegia was transferred by Mr Seddon to Mr Norman Capener at Plymouth with a suggestion that the spinal canal should be explored. Capener performed what he afterwards called lateral rhachotomy, removing not only rib and transverse process, as in Ménard's operation, but pedicle as well, thus providing an exposure of the canal; the cause of the spinal cord pressure was a spur projecting from the postero-inferior edge of a vertebral body, which was removed. Some years later G. L. Alexander and Norman Dott independently developed a similar operation. Antero-lateral decompression was a distinct advance and is the only safe procedure possible where the offending object is solid or semi-solid and contained within the spinal canal, whether it be massed granulation tissue, caseation, sequestrum, disc or bony spur.

Of surgeons whose work was intimately associated with country orthopaedic hospitals G. R. Girdlestone maintained an unremitting interest in the peculiar problems of the tuberculous cripple. He made many contributions to the subject, all of which he gathered up in his book of 1940 entitled *Tuberculosis of Bone and Joint*. It was more than a formal text-book in that the whole work was permeated with a personal experience at the Wingfield-Morris Orthopaedic Hospital for over twenty years. He advocated and described procedures for securing uninterrupted and adequate immobilisation but was disposed to venture upon radical operations earlier than most surgeons; osseous ankylosis by operation was for him both the curative and functional ideal. He was moved by the pathetic state of the paraplegic in spinal disease and proceeded to costo-transversectomy if recumbency tarried in its relief. His confidence in laminectomy was surprising, but that may well have been because antero-lateral decompression had hardly established itself; he welcomed it later.

Girdlestone in 1937 at Oxford had the distinction of appointment to the first chair of orthopaedic surgery in Britain, and his name is indissolubly linked with the Wingfield-Morris Orthopaedic Hospital which he founded. There he attracted men to watch his work and study the organisation of its regional clinics. He was a man of profound Christian belief and it was said there was something Franciscan in his affection for the poor and crippled. His friendship with Lord Nuffield was a potent influence in guiding that philanthropist's munificent medical benefactions.

Girdlestone received a training at Baschurch, whereas other early disciples of Sir Robert Jones were trained at Liverpool, of whom McMurray and Aitken come readily to mind. McMurray came from Queen's University, Belfast, to Liverpool where he remained for the rest of his professional career. After a period at the Southern Hospital he became private assistant to Sir Robert, whom he served with filial devotion. He was appointed surgeon to the Northern Hospital, and later became Director of Orthopaedic Studies and eventually Professor. With the instituting by Liverpool University of a higher degree in orthopaedic surgery McMurray threw himself with great energy into the building of a postgraduate school. Graduates came to it from all over the Commonwealth for teaching and training in orthopaedic surgery, proceeding later to the degree of M.Ch.(Orth.). McMurray, a tall, handsome and commanding figure, was unfaltering in his fidelity to the tradition of Thomas and Robert Jones; he leavened his forceful and dogmatic teaching with native humour and a paternal care for his pupils. He contributed greatly to the surgery of the hip by his displacement osteotomy of the femur which he published in 1935. This operation was an adaptation of the bifurcation of Lorenz for unreduced congenital dislocation of the hip. McMurray declared that the operation could be performed in a few minutes and shock was negligible. He was careful in the placing of lower fragment: " Oblique osteotomy is now made in shaft of femur passing from below upwards and inwards at such a level that the inner end of lower fragment lies about ½ inch below level of head of femur. Upper end of lower fragment is now displaced inwards and slightly upwards by means of a smooth lever and by abduction of leg so that shaft of femur comes to lie just below lower border of acetabulum." A plaster was retained for four to five months, during the last two of which the patient was allowed to get about weight bearing. When the plaster was discarded the patient in a few months regained comfortable movement at the hip. The operation was subsequently successfully adopted for non-union of a fracture of the neck of the femur.

McCrae Aitken migrated from Edinburgh to Liverpool and became one of the staunchest supporters of the Thomas tradition in orthopaedic surgery. In 1938 he wrote a valuable monograph entitled *Hugh Owen Thomas: His Principles and Practice* (Oxford University Press). On coming to London early in his career to pursue a higher qualification it caused him no little dismay to find that the caliper had no place in teaching hospitals in the treatment of a fracture or joint disease. He was about the first to be appointed to the military hospital at Shepherd's Bush, where his considerable experience was invaluable in demonstrating the use of the caliper and other Liverpool appliances in traumatic surgery and in teaching and training young officers from the United States and the Commonwealth. After the war he continued as surgeon to St Vincent's Orthopaedic Hospital and was appointed visiting surgeon at Oswestry, eventually succeeding Sir Robert Jones as surgical director. He devoted many years to the patient study of spinal deformity and wrote extensively about it. He was recognised by his colleagues as the best informed and the most successful amongst them in dealing with this difficult disability. He formulated a summary of his observations on the nature of spinal deformity and the principles which should guide treatment: " Scoliosis or rotato-lateral curvature of the spine, and kyphosis and lordosis or round and hollow back must all be taken in one group in any discussion of the deformities of the spinal column and trunk, for these several deformities blend and overlap to such an extent that they cannot be kept separate and distinct . . . Such deformity involves alterations in the relative positions of ribs and pelvis as well as of the spinal column, adaptive changes in muscles and ligaments and much the most important factor of all there are corresponding changes in that normal automatic reflex action of muscle taking part in maintaining the position of the body . . . At least two distinctive groups of reflex controlled muscles take part, namely the static group and the respiratory group. In the static reflex group must be included all the muscles of the lower limbs, pelvis and trunk which are concerned in keeping the body erect and balanced . . . In the respiratory reflex group are all the muscles concerned in maintaining respiration . . .

Disturbance of the proper reflex balance of the body is an essential factor which must always be kept in the forefront in considering the treatment of any case, whether an early mild case with little or no bony deformity or a later case with much distortion of the skeleton as well as alterations in the length and position of muscles. It is on this factor that the whole principle and practice of treatment by exercises or drill depends. Further, all correction of position by means of plaster (' forcible correction ') or manipulations is really directed towards shifting the centre of gravity of the trunk as a whole and straightening out the trunk so that the centres of gravity of the several sections of the body may be near enough to one vertical line for a re-education of the co-ordination of the 'postural' muscles to maintain this posture automatically, without conscious effort." In practice these principles were worked out by him in great detail. He passed through several phases in his attempt at correction by plaster jacket, ultimately deciding upon the application of a plaster that combined the essential ideas of Hoke, Haglund and Abbot with additions of his own. Although he made no claim " to have made a crooked vertebral column as straight as if it had never been distorted " he certainly did permanently improve the posture and well-being of very many individuals crippled with severe spinal deformity.

Among the countless casualties of the first world war were great numbers of peripheral nerve injuries, thus providing an unprecedented opportunity for the study of nerve regeneration. In this investigation there was happy cooperation between neurology and orthopaedics which was reflected in the Committee on Peripheral Nerve Injuries appointed by the Medical Research Council. Walter Rowley Bristow, who had displayed an intense interest in nerve and muscle injuries, was elected to this committee. During the war he cooperated with Sir Harry Platt in the study of nerve regeneration. The fruit of their investigation they communicated to the International Society of Surgery in 1923, in a valuable report on the remote results of operation for peripheral nerve injuries. They concluded that if the nerve ends could not be joined an autogenous cable graft should be used to bridge the gap. End to end suture in most patients produced an imperfect functional recovery; out of a large number of consecutive operations 20 per cent were complete failures. The extent of regained function after suturing varied with the particular nerve severed; the radial was head of the list; at least 50 per cent of patients with paralysis of this nerve had complete recovery after operation. Ulnar and median paralysis were disappointing. In the case of the ulnar nerve complete restoration of power in the intrinsic muscles of the hand was almost unknown; if recovery occurred it was invariably in the hypothenar muscles alone. But in spite of such indifferent ulnar nerve regeneration the hand often retained much of its usefulness except when finer coordinated movements of the fingers were required. With regard to the median nerve paralysis sensory recovery was always inadequate, causing considerable depreciation in manual function. Results of sciatic nerve suture were poor; a considerable number of paralysed lower limbs were ultimately amputated.

Rowley Bristow was appointed orthopaedic surgeon to St Thomas's Hospital in 1919. In a few years he organised and built up such a department that the number of patients requiring admission strained his allotted beds. Forced to look afield he made contact with an Anglican home for crippled children at Pyrford in Surrey; gradually this institution was transformed into a country orthopaedic hospital somewhat reminiscent of the evolution of Baschurch. There was something unique about this arrangement in that the Pyrford Hospital was not so much a country branch of St Thomas's as an extension of its orthopaedic department. Bristow's particular interest was directed towards traumatic orthopaedic surgery; he wrote on nerve and muscle injuries, internal derangement of the knee joint and fractures. He was a stimulating teacher of undergraduates, who crowded his clinic; he picked out essentials with clarity and emphasis, at the same time handling his patients with friendly understanding and good humour. Bristow was endowed with abounding vitality; a tireless worker with a zest for life and its friendships. As a tribute to his work at Pyrford the hospital has been named after him.

The growth of bone is of particular interest in skeletal surgery. It stimulated John Hunter to pursue experimental work on pigs which led him to declare that long bones were lengthened from the ends near the epiphysis and became thicker by deposition of new bone under the periosteum. Again bone was constantly undergoing change with simultaneous absorption and deposition of osseous tissue. Macewen first succeeded in grafting human bone from one individual to another and thereafter devoted many years to experimental investigation of bone growth, from which he concluded that the growth of a long bone occurred at the diaphysis, for he believed that the cartilaginous growth disc belonged to the diaphysis and not to the epiphysis. He also believed that periosteum had no osteogenetic power—it was purely a limiting membrane giving direction to bone growth but taking no active part in it.

In 1933 Professor H. A. Harris published a book of much originality entitled *Bone Growth in Health and Disease* (Oxford University Press), the result of experimental researches at the Institute of Anatomy at University College and University College Hospital. He noted transverse lines which cross the shafts of long bones, which he traced to arrested growth occurring during illness or disturbance of nutrition in childhood or early youth. Harris paid considerable attention to bone growth in deficiency diseases, with descriptions of histological and radiographic changes observed in proliferating cartilage, calcifying cartilage and bone; and he stressed how each process differed in its mutation. The osteoblast was a cartilage cell endowed with bone-forming properties. Growth at the end of a long bone was restricted to a mitotic annulus of dividing cartilage cells. Calcification he believed to be a biochemical process. He discovered that cartilage cells approaching the line of ossification contained glycogen which disappeared on conversion to bone. He suggested that glycogen was hydrolysed to hexose phosphoric esters which under the action of phosphatase and circulating calcium combined to form a deposit of insoluble calcium phosphate in the matrix.

RECENT TRENDS

The history of orthopaedic surgery is one of progressive evolution; for the first forty years it was concerned with the correction of congenital and paralytic deformities by splinting, manipulation and tenotomy. From about 1880 onwards the adoption of antisepsis permitted a bolder attack beginning first with osteotomy and advancing later to correction by open exposure. The war of 1914 and after quickened the pace. An answer for the insecurity of crippling was sought by joint stability through operation whether by tendon transplantation or arthrodesis or both. The open air conservative treatment of skeletal tuberculosis, with bone-graft surgery as a refinement, became the peculiar province of orthopaedic surgery and an increasing interest developed in the orthopaedics of trauma as a direct legacy of the war itself.

The story of the finding of penicillin and its development is already a romantic classic of science. The drifting of a mould spore on to Alexander Fleming's plate was as providential as the timely placing of him there with a mind already prepared to see its significance. " I found that while the crude culture fluid in which the mould had grown was strongly antibacterial it was non-toxic to animals and human leucocytes." It was fortunate too that Howard Florey was at that very time turning his attention to bacterial inhibition; " My own interest in the phenomenon of bacterial inhibition began in the 1920s." He, with Professor E. B. Chain and the rest of the Oxford team, succeeded in extracting a pure stable form of penicillin for human injection. Since that time the soil of the earth has been combed in many countries for other naturally occurring antibacterial substances. In 1944 Dr Selman Waksman of New Jersey isolated streptomycin from streptomyces griseus, a body antagonistic to the tubercle bacillus. These and other natural antibiotics together with synthetic sulphonamides introduced by Professor Domagk of Elberfeld in 1935 have revolutionised therapeutic practice.

The new therapy has had a very beneficial influence on orthopaedic surgery. The lurking

threat of sepsis has been greatly mitigated, protection has been given to operative procedures beyond what was possible by surface antiseptics alone. Bone infection such as osteomyelitis is much more readily controlled and the compound fracture is less menacing to life and limb. Chemotherapy in joint tuberculosis retards the destructive process of the disease, shortens the duration of the malady and permits of an earlier fusion operation. When invasion of a joint is comparatively early, with no sign of bone erosion, rigid immobilisation may be relaxed and joint movement permitted under supervision.

There is change in the incidence of disease. Rickets has all but vanished; on its deformities osteotomy at one time flourished. Wonderful to relate, tuberculosis at long last is a declining disease due to protective and improved curative therapy and social betterment with its clean milk, better feeding and education. Infantile paralysis is still a dread menace, but there is good reason for believing that a safe protective vaccine will soon emerge.

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RICHARD WATSON (1737-1816)

Bishop of Llandaff, Hunterian Trustee

A Chapter in the History of Orthopaedics

SIR HARRY PLATT, MANCHESTER, ENGLAND

President of the Royal College of Surgeons of England

On December 3, 1799, at a special meeting of the Court of Assistants, the Company of Surgeons of London, soon to become the Royal College of Surgeons of England, accepted the custody of John Hunter's famous collection which had been purchased by the Government for £15,000. One of the conditions laid down by Parliament was that there should be a Board of Trustees thirty in number, fourteen of whom were to be appointed by the Lords of the Treasury. In the first list of Government nominees there appears the name of Dr Watson, Bishop of Llandaff. There is no recorded evidence that Dr Watson took his Hunterian duties seriously, for he never attended a meeting of the Trustees. It might therefore be assumed that nothing in the life of Bishop Watson touched in any way the life of the Royal College of Surgeons either in his own day or in the century and a half which lay ahead. And yet by one of his actions he was to influence profoundly the organisation of orthopaedic surgery in the second half of the twentieth century.

Richard Watson was a figure of some importance in English public life in the latter part of the eighteenth century (Fig. 1). He was a Westmorland man, born at Heversham in 1737 and educated at Heversham School, of which his father was headmaster for nearly forty years. The most distinguished pupil of that school was Ephraim Chambers, the author of the celebrated dictionary.

Watson went up to Cambridge at the age of sixteen, supported by an exhibition attached to the school. He was admitted to Trinity College as a Sizar, but after some two and a half years was awarded a scholarship by the master. He read both classics and mathematics and his facility in the latter subject was such that he coached men in the year below him. He took his B.A. degree in 1759, appearing in the list as Second Wrangler. In actual fact the examiners had placed him first but the leading Moderator put one of his private pupils and a student of his own college—St John's—as Senior Wrangler! In 1760 Watson became a Fellow of Trinity and shortly afterwards refused two offers of appointments in the Church—a curacy at Clermont and a chaplaincy in India. Two years later he became a Master of Arts, a Moderator for his college, and now looked round for other fields to conquer. He had not long to wait. In 1764 the chair of chemistry in the university became vacant and Watson decided to offer himself as a candidate despite the fact that he knew nothing at all about the subject. He had never read a word nor seen a single experiment. His only rival for the chair, an eminent London physician, withdrew from the contest when told that Watson intended to deliver a course of chemical lectures in the university—evidently an unheard of innovation. The senate duly elected Watson, who at once sent to Paris for what he called an " operator "—a laboratory technician, no doubt. For the next fourteen months he buried himself in his laboratory for a considerable part of each day, and at the end of this period of self-education delivered a course of lectures " to a very full audience, consisting of persons of all ages and degrees." Two years later he delivered another course. When appointed to the chair, Watson found that there was no stipend attached to the office, and the Department of Chemistry consisted of one room. He therefore applied through the Chancellor of the University, the Duke of Newcastle, to the First Lord of the Treasury, then Lord Rockingham, for a Crown subsidy of £100 per annum.

This request was granted and it was not long before the chairs of anatomy, botany, and common law were similarly rewarded. In 1768 Watson composed and printed a treatise embodying his lecture course and was elected a Fellow of the Royal Society after submitting a thesis on solutions of salts.

During his active period as a teacher of chemistry he was seized with a desire to perfect his knowledge of human anatomy. A body was obtained from London, which he dissected in his Cambridge laboratory with the assistance of two professorial colleagues, one a mathematician and the other a theologian who afterwards became an Irish bishop. The remains of the dissected body were placed in a box which was given to an old soldier with orders to carry out a decent burial. For some unknown reason the box was thrown into the River Cam and a few days later parts of the corpse drifted ashore and led to the suspicion that a murder had been committed.

FIG. 1
Richard Watson, Bishop of Llandaff, Hunterian Trustee

In his earlier years as a Cambridge don and professor, Watson played an important part in the general academic life of the university. He soon showed himself to be a reformer in education. When holding the office of moderator he addressed the senate in Latin, pointing out the defects in the educational system at Cambridge, where at that time noblemen, fellows and commoners attended very few classes. Watson recommended the institution of annual public examinations, the use of set books, an increase in the number of examiners, and the recognition of a distinction between the ordinary pass and an honours degree. Such reforms were rejected by a narrow majority of the senate in 1774.

In 1771 the Regius Professor of Divinity died, and Watson at once saw another opportunity for advancement in life. His qualifications in academic theology were non-existent; he was neither a Bachelor nor a Doctor of Divinity but he had seven days' grace before the election. By dint of much travelling and lobbying he obtained a royal mandate to present himself for the D.D. on the day before the examination which consisted of written essays in Latin read as a dissertation. At the age of thirty-four the ambitious Richard Watson was appointed

to the Chair of Divinity, to which was then attached the annual stipend of £300. With a modest but secure income he soon felt able to enter the holy state of matrimony, choosing his wife from the Lakeland countryside (1773). The day after his marriage he took a sinecure rectory in North Wales, a living in the gift of the Duke of Grafton which he soon exchanged for a prebend in the church at Ely.

Although Watson was well aware that advancement in the eighteenth century usually depended on securing the right kind of patronage, he was a man of independent views and was not afraid of incurring criticism. In politics he was a Whig and supported the movement for parliamentary electoral reforms in Cambridgeshire. In the affairs of the Church he was a libertarian and urged Lord Rockingham to reform the Bench of Bishops by equalising their incomes. In 1776 Watson preached the Restoration and Accession Sermons before the university, published later under the title of *Principles of the Revolution Vindicated.* This lost him the support of certain people who had intended to suggest him for the vacant post of provost at Trinity College, Dublin. The sermons also aroused the disapproval of the Archbishop of Canterbury, which led to an exchange of letters between the primate and himself.

Watson's independent attitude must have impressed some people in high places, for in 1782 he was informed by the Duke of Grafton that the see of Llandaff was about to become vacant and that his name was to be submitted to the new First Lord of the Treasury, Lord Shelburne. In July of that year he had an interview with the Minister and his appointment to the see was confirmed on May 30, 1783. Now in his forty-seventh year, Watson made his first speech in the House of Lords on the evils of simony. From Westmorland to Westminster via Cambridge; Student and Fellow of Trinity; Professor of Chemistry; Regius Professor of Divinity; Bishop of Llandaff—he had come a long way.

During the next twenty years, in the House of Lords and in his publications, Richard Watson supported many unpopular causes—amongst them Irish Independence and the Independence of the American Colonies. He had already expressed publicly his disapproval of Pitt's Commercial Treaty with France as likely to ruin England's trade. He was also a critic of William Pitt's management of the war with France. In a letter to the Prime Minister in 1797 he advocated a capital levy of 20 per cent, but Pitt's reaction to this was to impose an income tax! Watson returned to the attack in the House of Lords in 1804, urging the need for the conscription of men, the requisition of arms and horses, and once again a capital levy. In 1787 he had been consulted by the Government on the manufacture of gunpowder and had suggested a method which he claimed had saved the country £100,000 a year. For this service he received no special recognition. During this year he contracted a severe attack of dysentery, and his slow recovery prompted a letter to Mr Pitt asking for his release from the Chair of Divinity. The immediate reply was evasive. It was said that Pitt suspected Richard Watson of desiring to succeed to the see of Durham, the emoluments of which were then £5,000 a year, a princely stipend when compared with the £500 a year of the see of Llandaff. But Watson, though himself a pluralist, had never abandoned his advocacy of the equalisation of incomes amongst the bishops and the establishment of a minimum basic income of £100 a year for all parish clergy. He was moreover now in possession of ample private means, for during the previous year his close friend Mr Luther, living in Essex, had died leaving him an estate which Watson sold some time later to Lord Egremont for £23,000. This windfall enabled him to provide for the future of his family.

During his convalescence from the attack of dysentery he found at Calgarth on the shores of Lake Windermere a site for his future home. In 1789 he laid the foundation of his modest mansion. He had said farewell to Cambridge, and now from Calgarth, where he spent eight months in the year, planting trees, enclosing wastes, and engaging in farming, he sallied forth on visits to London to speak in the House of Lords, or to tour his diocese.

Richard Watson was the first of the intellectuals to make his permanent home in the

Lake District, and at Calgarth he often entertained the poets and authors who in later years had settled there. At Elleray a mile away, there was Christopher North, essayist and philosopher long identified with Blackwood; at Old Brathay, Charles Lloyd; and at Grasmere eight miles away, Wordsworth and De Quincey. Southey was at Keswick on Derwentwater, and Coleridge lived for spells at Keswick and Grasmere. We can imagine the lively conversations amidst scenes of great beauty.

Although his life at Calgarth was one of absorbing interest and happiness, Richard Watson's desire for promotion on the Bench of Bishops still lingered. What he resented most of all was to see his juniors translated to the most important dioceses, and by influences which he despised. In the year of his first breakdown in health a letter from the Duke of Norfolk containing news of the death of the Bishop of Carlisle followed him from Cambridge to Westmorland. The vacant see however was offered by Pitt to a relative of Sir James Lowther—a piece of pure jobbery in Watson's opinion. In his seventieth year, when an

FIG. 2
Calgarth—Early twentieth century.

uprooting would have been a disturbing event, he was disappointed not to be offered the vacant see of St Asaph.

In 1811 Watson sustained a paralytic stroke and lost the use of his right hand, though retaining his general bodily vigour. Two years later his health began to decline, but he lingered on for another three years and died on July 4, 1816, still Bishop of Llandaff and a Hunterian Trustee.

Richard Watson's eighteenth century mansion in its estate of some seventy acres remained in the possession of his heirs and successors throughout the nineteenth century. During that period the house was enlarged by the addition of wings (Fig. 2). A few years before the outbreak of the first world war the estate was bought by Mr Oswald Hedley, who in November 1914 handed over the mansion to be used as a convalescent hospital for Belgian wounded. Mrs Hedley, who had taken a leading part in the organisation and running of the hospital, died in 1916 before her idea of remodelling the hospital as a rehabilitation centre for wounded British officers could be carried out. After her death Calgarth was closed for a little more

than a year. During this time Mr Hedley gave carte blanche for its reconstruction and equipment, and on its reopening towards the end of 1917 dedicated it to her memory.

The Ethel Hedley Hospital, as it was now called, became an auxiliary unit of the military base hospital in Manchester, the Second Western General Hospital, from which its patients were drawn. In common with all auxiliary hospitals it was visited from the base hospital once a month, and this duty was laid on a young surgeon holding the rank of Captain in the R.A.M.C.T. who had been placed in charge of the military orthopaedic section of the Second Western General by Sir Robert Jones. By 1919 the flow of military patients had dwindled almost to vanishing point, although for a short time the beds were occupied by pensioners, at first ex-officers and later other ranks. The large capital outlay on the reconstruction and the

FIG. 3
O. W. E. Hedley, Esq., O.B.E.

expense of maintenance of the Ethel Hedley Hospital had been borne entirely by Mr Hedley (Fig. 3). It is known that he must have spent more than £100,000 on the hospital during the war years. The mansion was no longer suitable as a private residence; its reconversion for that purpose would have been a most expensive undertaking. What was to become of the hospital? As a result of extensive experience in the reconstructive surgery of the locomotor system arising out of the war, the art and science of orthopaedic surgery had undergone a renaissance. Sir Robert Jones and that remarkable woman, Agnes Hunt, were devising orthopaedic schemes for the civilian population; the Oswestry pattern was taking shape—a central orthopaedic hospital served by a chain of after-care clinics distributed widely throughout the rural areas. The needs of the crippled child were first and foremost in the

minds of Sir Robert and his small band of younger disciples. In the Lakeland counties of Westmorland, Cumberland, and Lancashire north of the sands of Morecambe Bay, with a population of nearly 400,000, the need for such an organisation was obvious. A number of after-care clinics had already been established in Cumberland under the guidance of Dr Kenneth Fraser, County Medical Officer of Health, but there was no central long-term hospital; beds were leased in hospitals remote from Lakeland.

All interested eyes turned to the Ethel Hedley Hospital. With some diffidence Mr Hedley was approached once more. With princely generosity he rose to the occasion and the third reconstruction of Calgarth was undertaken. It now became a children's orthopaedic hospital equipped for active surgical treatment, and when opened in 1920 was recognised by the

FIG. 4
The Ethel Hedley Hospital, Windermere; high summer 1955.

Board of Education as a special hospital school for the combined education and treatment of physically handicapped children in fulfilment of the provisions of the Fisher Education Act of 1918. At first the in-patient accommodation consisted of twenty beds, but in 1924 this was increased to fifty beds which necessitated the building of a new wing containing plaster rooms, gymnasium, physiotherapy rooms, and quarters for the nursing staff. During the past thirty-five years the Ethel Hedley Hospital, once the home of Bishop Watson, Hunterian Trustee, has undergone only minor alterations in internal structure (Figs. 4 and 5).

We may look behind the scenes and take a glimpse of one whose influence as the guide, philosopher and friend of Mr Oswald Hedley was ultimately responsible for the transformation of Calgarth Park into its present-day form. Charles Henry Hough was born in 1855, the son of Dr James Hough of Cambridge who was elected a Fellow of the Royal College of

Surgeons under the twenty-year rule. Charles Henry Hough was educated at Uppingham under Thring, was an undergraduate at Cambridge, and received his clinical training at St Thomas's Hospital where he became an M.R.C.S. in 1878. After qualification he held the post of house surgeon at the Derby Royal Infirmary and later settled in that city, combining general practice with surgery as a member of the honorary staff of the hospital. Hough practised in Derby for a quarter of a century, becoming senior surgeon at the hospital, and, although always a general practitioner, was consulted freely by his professional brethren in the city and county. In 1903 he left Derby for Westmorland and at the northern end of Lake Windermere built a house on the slopes of Lough Rigg in sight of Brathay Hall, the family

FIG. 5
The Ethel Hedley Hospital, Windermere; high summer 1955. Lake Windermere in the background.

home of his wife, who had only one short year to enjoy her new home on the rock before her death. Here Charles Henry Hough began the construction of a rock garden which became and remains one of the most beautiful of its kind in Great Britain, with plants brought from many lands, from the mountain slopes of Switzerland and from far off Tibet (Fig. 6). It was not a retirement for Hough, for he had resumed practice in Ambleside and once again became much in demand as a general consultant.

On the outbreak of the 1914 war, at the request of Mr Hedley, Hough gave up his practice in order to become Medical Superintendent of the Calgarth Hospital, and he continued in this office throughout the subsequent reconstructions and changes in its function. In its earlier years as a children's orthopaedic hospital he assisted at operations, visited after-care clinics and published its first report—an account of the work done during the first ten years (1920–1930). Six months before his death in 1933 he was elected a Fellow of the Royal College

of Surgeons as a member of over twenty years' standing. It was an honour richly deserved, and for him a moving experience when he entered the council chamber to be welcomed by Lord Moynihan in his most felicitous mood.

It has been my great privilege to act as surgical director of the Ethel Hedley Hospital since its opening thirty-five years ago. To escape once a month from the turmoil of the life of a surgeon in a great city; to spend a Saturday amidst scenes of great beauty, not resting in a physical sense, but enjoying a full working day comprising ward round, operations, and out-patient consultations, has been a constant source of inspiration and joy. In my earlier years I used to stay at White Craggs, the house on the rock; and in the spring and summer

FIG. 6
C. H. Hough on the rock.

months would walk to the top of the rock garden before proceeding to the hospital for the day's work. High up on the rock there is now a shrine which commemorates the life of Charles Henry Hough; his brother James Hough, once Bishop of Woolwich; his cousin Cecil Boutflower, once Bishop of Tokio and later of Southampton; and his eldest grandson, killed in the last war.

And thus we see how Richard Watson, Bishop of Llandaff and Hunterian Trustee, built more than he knew; and how the whirligig of time has revealed a chain of events which has linked his day with the age in which we live. Richard Watson; Oswald Hedley; Charles Henry Hough; and the President of the Royal College of Surgeons and a custodian of the Hunterian Collection in the year of grace 1955. Such is the story of an eighteenth century Westmorland mansion.

SOME OBSERVATIONS ON CONGENITAL DISLOCATION OF THE HIP

BRYAN MCFARLAND, LIVERPOOL, ENGLAND

The entity passing under the name of congenital dislocation of the hip is so complex that the sense of privilege in contributing to this special issue of the Journal in honour of Sir Thomas Fairbank does not completely offset a feeling of awe engendered not only by the occasion but also by the vastness of the subject. With a similar feeling Isaak Walton must have approached his task of writing the *COMPLEAT ANGLER*. In a letter of dedication to his patron, John Offley, he said " Sir, this pleasant curiosity of fish and fishing, of which you are so great a master, has been thought worthy the pens and practices of divers in other nations that have been thought men of great learning and wisdom; . . . I do freely confess that I should rather excuse myself than censure others, my own discourse being liable to so many exceptions against which you, Sir, might make this one, that it can contribute nothing to your knowledge." At the opening of his address to his readers, Walton said: " I think fit to tell these following truths: that I did neither undertake nor write nor publish and much less own this Discourse to please myself; and having been too easily drawn to do all to please others, as I propose not the gaining of credit by this undertaking, so I would not willingly lose any part of that to which I had just title before I began it, and do therefore desire and hope if I deserve not commendation yet I may obtain pardon." Isaak eased his way through an ambitious project by introducing a novice *Venator* who attached himself as a *Scholar* to the expert angler *Piscator*. At appropriate intervals the novice asked a question of his chief in order to clarify a point or to change the theme.

A similar device might ease my own way in this discourse on some aspects of congenital dislocation of the hip. May we imagine that a senior surgeon of experience—*Mentor*—is talking to an enquiring student who has the good sense to be interested in orthopaedics.

Mentor—In the time available to us we cannot deal with the whole of this subject, so we must select parts. It seems appropriate to discuss those aspects in which knowledge has advanced, or in which understanding has increased since orthopaedic surgery was recognised as a speciality. Just over thirty years ago Vittorio Putti, Robert Jones and Thomas Fairbank were teaching and training young men in the treatment of congenital dislocation of the hip against a background of the work of Lorenz, Schanz and Codivilla.

We need spend little time on etiology. Controversy is still active between the geneticists and the environmentalists, though certain facts of geographical and racial distribution seem to favour the geneticists. Nevertheless their explanation why usually girls are stricken is not altogether satisfying; and the experimental work on the artificial induction of congenital defects cannot be ignored. It may be that a combination of both factors ought to receive more attention, because possibly there is a genetic flaw which may be boosted by adventitious factors. Such a flaw might affect one or other or both major components of the hip joint and operate in widely varying degrees. Thus at one extreme there would be gross deficiency of the pelvic and femoral components, and at the other a deficiency so slight as to be revealed only as growth continued or by the stresses of advancing age.

Consider this case of absence of one half of the pelvis (Fig. 1) and this example of absence of the upper three-quarters of the femur (Fig. 2): this bilateral coxa vara with gross deficiency of the neck of one femur, and a relatively mild aberration of growth in the other (Fig. 3): or this bilateral dysplasic dislocation of the hip (Fig. 4): and compare them with this mild dysplasia of both hips in which osteoarthritis is only now beginning at the age of thirty-five, in a patient hitherto untroubled (Fig. 5). A case is recorded of a patient seemingly normal

FIG. 1

FIG. 2

Figure 1—Congenital absence of half the pelvis. Figure 2—Congenital absence of the upper three-quarters of the femur.

FIG. 3

FIG. 4

Figure 3—Bilateral congenital coxa vara: gross defect in the neck of one femur and aberration of growth in the other. Figure 4—Gross bilateral dysplasic congenital dislocation of the hip.

FIG. 5

Woman aged thirty-nine years. Dysplasia of both hips. Increasing pain in the right hip during the last five years.

at birth in whom during the subsequent five years defects developed leading to coxa vara. We might consider the possibility of a common basis for developmental affections of the pelvis and femur, including congenital dislocation of the hip.

Student—These are deep waters. It would be interesting to turn to something more tangible. For example, what is the best treatment for congenital dislocation of the hip?

Mentor—A simple answer to this question is quite impossible. It is necessary to know the age of the patient as well as the type or stage of the dislocation, and whether it involved one hip or both. Details of treatment have changed many times but the objective and the principles remain unaltered. The objective to be gained is normal function. The principles are to secure reduction as early as possible by the simplest and most gentle means, and to maintain it until growth has made it lasting.

Granted these principles, the detail of any particular pattern of treatment is less important than the average age at which it is begun. In 1925 and for some years after that the usual age at which children were brought for treatment was about three and a half years: today the average age is much nearer twelve months. It is now well recognised that few perfect hips can be achieved in any large number of children first treated at the age of about three years—certainly not much more than 40 per cent; whereas if treatment is begun at the age of about twelve months this figure should reach 80 to 90 per cent. The importance of being early is clearly paramount. The age when treatment begins will depend upon the dislocation first being suspected; suspicion leads in turn to investigation, diagnosis and treatment.

It has been said that if every new-born baby were examined for the possibility of congenital dislocation of the hip, treatment would always start within a few months of birth. It has also been said that limitation of abduction of the hip at this age is significant, and that radiographs will reveal dislocation whenever present. There are fallacies in these statements. Limitation of abduction may be difficult to determine in a little baby and it may be present in conditions other than congenital dislocation of the hip—for example in arthrogryposis, spastic hemiplegia or paraplegia, and congenital coxa vara. On the other hand, and for obvious reasons this is the more important, it may not be present in congenital subluxation or the milder degrees of dislocation. Moreover radiographs may not be capable of exact interpretation even by most experienced observers. I well remember hearing Putti give an account of the earliest diagnosis on record. Because of the prevalence of congenital dislocation in her part of northern Italy a mother sent her baby only twelve hours after delivery to the Rizzoli Institute: she feared that one hip might be dislocated. Clinical and radiographic examination suggested that the hip was indeed dislocated but Putti said that he could not be sure. The mother, anxious to suppose her child normal, seized on this uncertainty and declined treatment. A year later she brought the child for examination again and there was then no doubt.

Student—Is then the check to abduction movement of no value in diagnosis?

Mentor—Certainly it is of value, but a feeling of this check to abduction is only one of the straws to show which way the wind blows. Another is the unusual appearance of the leg, which seems to be slightly externally rotated: yet another is elevation or duplication of the crural crease; and when the child kicks as babies do, the affected side tends to circumduct in flexion. There may be asymmetry when there is dislocation of only one hip, but not of course when both joints are displaced. It may sometimes be possible to elicit telescoping, but at such an age this is not fully reliable, especially when the joint is only subluxated. Fortunately a decison need not rest upon one examination alone. The prudent course is to arrange for re-examination after a time long enough for the signs to become more definite—but short enough not to prejudice the result. If there is doubt, a few months of treatment in abduction is better than further temporisation with the danger of missing the opportunity of a complete cure.

Student—Accepting that the age of detection affects the outlook, could you now deal with treatment?

Mentor—I would prefer first to indicate the types of dislocation. These have become more clearly differentiated during the period we are considering. Then we shall have in our minds pictures of the different types, and we can see that not only the age but also the type of displacement affects the treatment we may employ, as well as the result that may be expected. We must of course remember that the more severe displacements of the hip are usually recognised at an earlier age; but even so it is doubtful whether this offsets the difficulties of creating a normal joint. The lesser displacements, though more easily restorable to normal, tend to escape notice; and therefore unless some collective action is undertaken by the community the best chances of ultimately securing a very high proportion of normal hips may continue to be lost.

FIG. 6
Baby girl. Subluxation or " pre-dislocation " of left hip.

Student—Do you then recognise two types of dislocation—the mild and the severe?

Mentor—The fact of the matter is that I recognise three types: the good hip with mild displacement, the good hip with severe displacement, and the frankly dysplasic hip. By mild displacement I mean a dislocation in which the abnormality is less than minimal, if such a thing is possible—in other words so slight that one may have to act almost on suspicion. In the typical case the child will be not more than a few months of age, and attention will have been attracted by one of the signs already mentioned. Radiographic examination may have shown that the ossific nuclei of the femoral heads are not present on either side, or if present they are unequal. and the femoral shafts may be slightly shifted laterally, and there is a defect in the upper lip of the acetabulum, perhaps too slight even to raise the acetabular index. Such a hip is only in a position of subluxation, or as it has been called " pre-dislocation " (Fig. 6).

The second type is that in which all the classical signs are present. Of course they become more noticeable as time passes. It seems likely that whereas the very mild type to which I have already referred if neglected becomes the classical subluxation, this second or moderate type of displacement is, in fact, a dislocation from the beginning. Certainly the signs are present at a very early age. Probably the two most clinically reliable signs are the shift upwards and outwards of the leg and the telescoping. You will appreciate the difficulty of confirming other signs, such as shortening, elevation of the trochanter, and dorsal displacement of the femoral head, which are either uncertain or difficult to elicit in small children who do not tolerate long examinations. Radiographs show that there is some displacement of the femoral head; the capital epiphysis is small; the shift upwards and outwards is moderate; the acetabular lip is deficient but not markedly so; and the neck of the femur is at a reasonable angle to the shaft (Figs. 7*a* and 7*b*).

In the third type all the classical signs are well defined. The asymmetry in unilateral dislocations, the abnormal position of the legs in relation to the body in bilateral dislocations, the alteration of movement, the hollow in Scarpa's triangle, the gluteal prominence—all these are very obvious. Radiographically the changes are much more than an exaggeration of the second type. The head of the femur is well up on the dorsum ilii, and the capital nucleus is undersized or in earlier cases may not be showing at all. Even if the condition is bilateral, retardation of growth of the capital epiphysis will still be quite obvious. Moreover the defect in the upper part of the acetabulum may be so great that the slope from the depth of the

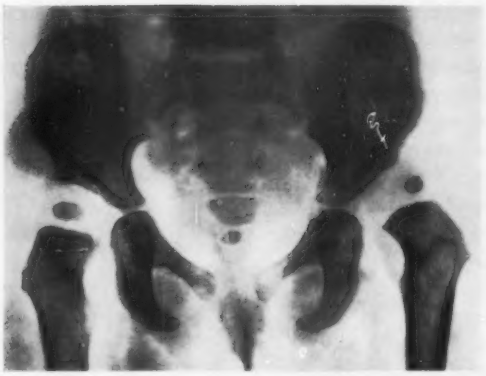

FIG. 7*a*

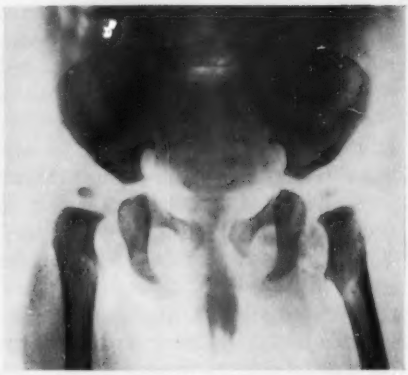

FIG. 7*b*

Figure 7*a*—Girl aged eighteen months. Second type, *i.e.*, " the good hip severely displaced." Left side.
Figure 7*b*—Example of same type: bilateral dislocation.

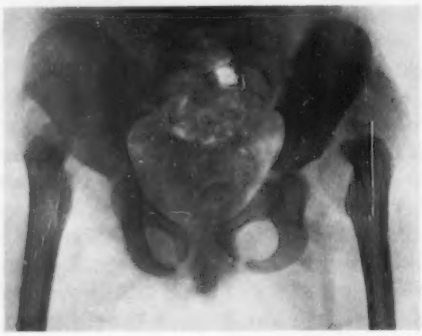

FIG. 8
Girl aged two years. Third type: " the frankly dysplasic hip." Bilateral.

acetabulum to the blade of the ilium is almost uninterrupted: the angle between the neck and shaft of the femur is minimal: the false acetabulum is apparent at an early stage (Fig. 8). I suggest that this last type is basically different from the first two types, and that such dysplasia exists embryonically rather than foetally. It is interesting to note that the defect of the acetabulum involves mainly the ilium, and that the defect spreads beyond the acetabulum proper. Look at this tracing of the radiographs of a child aged eleven months who unfortunately died shortly afterwards of epidemic enteritis (Fig. 9). Incidentally the angle of anteversion in the dislocated hip was only 5 degrees greater than in the normal hip (Fig. 10). This water-colour drawing is of the dissected pelvis of that child (Fig. 11). It will be seen that the acetabular defect involves mainly the iliac segment; it is gross, for instance the anterior inferior iliac spine is absent; and although the child had never walked the capsule had already " travelled " upwards, the ligamentum teres being several times its normal length.

Student—May we now come to treatment?

Mentor—It is a pleasant relief to turn to treatment. It is clear that the first type, if it is detected early, can be treated very simply and that a normal hip should be restored. Retention in abduction by one of the well known frames or any similar apparatus is quite enough. Care must of course be taken that the head is properly centred in the acetabulum.

In the second type, which I have suggested may arise in foetal life, treatment has to be

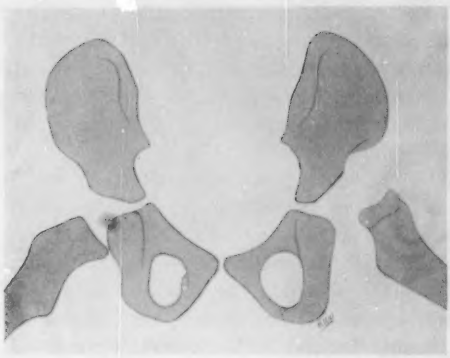

FIG. 9

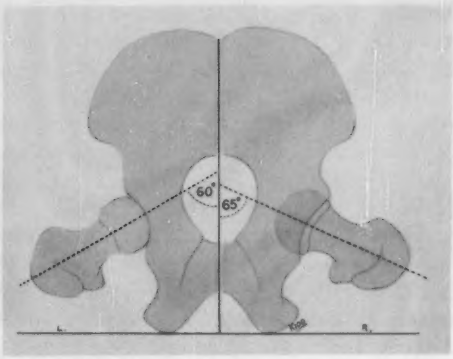

FIG. 10

Figure 9—Tracing of radiographs of a child aged eleven months who died shortly afterwards of epidemic enteritis. Figure 10—Diagrammatic drawing of dissected pelvis of the same child showing the very slight difference in the degrees of anteversion.

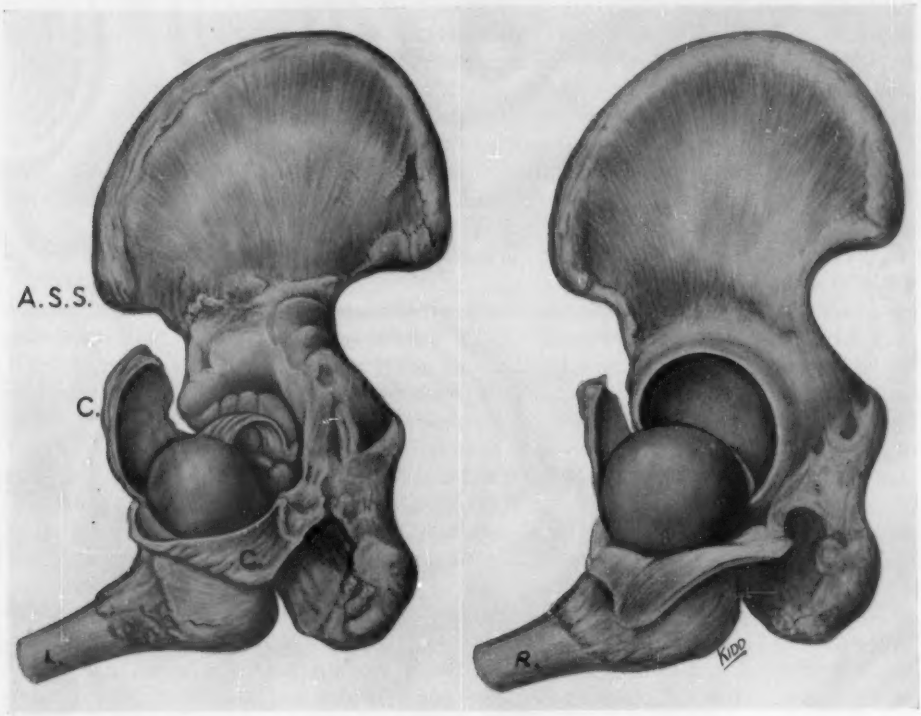

FIG. 11

Water-colour drawing of dissected pelvis of the same child aged eleven months. In addition to the defects mentioned in the text, note the adventitious fat in the base of the acetabulum. The capsule " C " has been divided and retracted. In order to facilitate comparison a mirror image of the right (normal) hip is shown.

modified according to the age at which it is commenced. Up to about the age of eighteen months, closed manipulative reduction is simple, and easy, and the prognosis is good. The technique of reduction has not changed in the last thirty years. The hip is reduced with the thigh in the position of flexion to a right angle and in slight adduction. The surgeon applies traction with one hand to the thigh, and with the other maintains pressure over the trochanter, thus easing the head gently into the acetabulum, the final move home being accompanied by a few degrees of internal rotation of the femur.

The method by which the head is retained in its proper position is a matter of individual preference, but one or two points should be emphasised. On no account should immobilisation involve undue stress. It must be remembered that while the hip has been dislocated the main adductor and flexor muscles have assumed a length which is too little when the dislocation is reduced. Thus tension is caused when abduction is forced. The resulting tension is translated into pressure by the head of the femur against the acetabulum, and reaches its maximum when abduction is at a right angle to the mid-line and the thigh lies in the coronal plane. This is unfortunately regarded as the " frog position " and may well be a significant factor in the onset of osteochondritis of the femoral head. If the thigh is a few degrees above the right angle, and if the thigh is similarly a few degrees in front of the coronal plane, the pressure is not excessive. This point at which the head is quite safe and not subjected to excessive tension can easily be appreciated by the operator's hand. Moreover in this position the capsule is relatively lax; whereas in what is sometimes regarded as the frog position (but which I think is an exaggeration of the original " frog ") and also in a position of forced abduction and internal rotation, the capsule is either tense, or twisted in such a way as to occlude its blood vessels. In my opinion osteochondritis of the femoral head is more likely to be the consequence of occlusion of the vessels by torsion or tension of the capsule than of direct pressure on the head itself. This strangling effect may occur not only at the time of the first manipulative reduction and in the first position of immobilisation, but also at the first change of position when the limb is replastered, especially if this is done abruptly or with any degree of force. The change of position at various stages of treatment must be so gradual as to allow time for adjustment of the capsule to the new position.

Student—Is there not a movement towards tenotomising the adductors with a view to minimising the stress in the hip?

Mentor—Yes, and this is intriguing because it seems possible so to achieve stability with only a moderate degree of abduction. But I wonder why this tenotomy was abandoned thirty years ago after it had enjoyed such a long period of popularity. As a matter of fact it was just going out when I was coming in. Probably the question might repay re-examination. In the meantime a tendency is developing which appears very promising. This is the practice of avoiding prolonged recumbency. The child is first rigidly protected with the legs nearly in the coronal plane and is able thus to crawl about. At a relatively early stage movements of the hip joints, still with the legs prevented from moving forward from the coronal plane, are permitted and encouraged. There seems little doubt that although the total period of protection cannot be materially reduced because it must be provided until growth has reached a certain stage, nevertheless the difficult period of varying length between full immobilisation and full activity, which the older regime involves, is rendered easier and shorter, and in fact may be avoided altogether.

It is obvious that in certain cases of this second type of dislocation the pattern of treatment may be varied. For instance such hips have been reduced and retained in good position without any manipulative reduction under anaesthesia, the gradual traction and the retention being provided by a Thomas's splint. I remember during the second world war, when it was difficult to provide in-patient treatment, that some such children were treated thus as out-patients (Figs. 12 to 17). But it is doubtful whether it is applicable in general, and with constantly changing nursing staff it is also doubtful whether it is safe as a routine.

FIG. 12

FIG. 13

Figure 12—Girl aged eighteen months. Left congenital dislocation of hip. Second type. Figure 13—Same patient as Figure 12, two months later. Treated as an out-patient. Dislocation reduced by traction on Thomas's splint.

FIG. 14

FIG. 15

Figure 14—The same patient four months later. Note commencing formation of acetabular roof. Figure 15—The same patient six months later, *i.e.*, after twelve months' treatment (as an out-patient). Note increasing growth of acetabular roof.

FIG. 16

FIG. 17

Figure 16—The same patient three months later, *i.e.*, after fifteen months' treatment. Marked improvement in the acetabular roof. Figure 17—Two years later, *i.e.*, after three and a quarter years from commencement of treatment. Acetabular formation practically normal.

Student—All this that you have just said is on the assumption that the patient was very young and that the dislocation was of the second or foetal type—that is to say, definite though not marked and severe. Supposing that the child were older, say two and a half or three years, or that the dislocation were of a rather more severe grade, would you expect in such a case to obtain reduction quite easily in the way that you have described?

Mentor—At the first attempt, no. It must be remembered that the manipulation has to be gentle. On no account must force be used. If without force the hip cannot be reduced, the attempt should be abandoned and traction should be applied to the leg for about fourteen days. A second attempt at reduction may then be successful. But if this is still not possible without using force, the same procedure should be repeated after a second fourteen days. Even then reduction may be unsuccessful but one would have gained from the manipulations (and the radiographs) a picture of the dislocation and what is preventing reduction. I would then undertake an open operation and would expect to find that complete reduction was prevented either by a large fold of capsule or by a pad of adventitious fat. At such a stage one would not find what one does find later—namely that the acetabulum is not of an appropriate size or shape to receive the head. Even when open operation is performed it is necessary for the head to fit into the acetabulum without undue pressure between the two.

Student—Would you not have taken an arthrogram?

Mentor—No. I am not able to learn enough from such a procedure to justify its use. The shadows are too variable to indicate more to me than can be learned from good plain radiographs and stereoscopic views, and from my hands when attempting to reduce the hip. By learning from the hands I mean that it is possible to know whether the head can be made to reach the acetabulum, whether the acetabular rim is well or poorly developed, and whether there is some obstruction inside the acetabulum.

Student—You have made no mention of constriction of the capsule to which I think the description " hour-glass " has been applied.

Mentor—I do not believe that such a constriction commonly exists. I have opened a great many hip joints but I have never seen a true hour-glass constriction. I think the appearance of a constriction could be caused by twisting the capsule in rotating the leg. I think also that the lower part of the capsule can be drawn across the acetabulum in the same way as a small bag attached to the rim of a golf-hole, for instance, would, if drawn to one side, occlude the hole. (And I must say when one is playing golf it sometimes seems as if there were an invisible bag drawn across the mouth of the hole.) It might be thought that the constriction has not been recognised because of the small size of the structures in childhood and because the tissues are soft and blended one with the other. But if the constriction is there in childhood it would persist into adult life. My experience with the congenitally dislocated hips of adults has been that the capsule is loose and voluminous—there is certainly no constriction.

There remains the third type, which we have called the severely dysplasic or embryonic type, with appearances approaching those seen in arthrogryposis. If such a case is seen very early, it will yield to the measures outlined above. But if it is seen later, at three, or two, or even one and a half years of age, it may present a very difficult problem. This is because not only is the acetabulum in its upper part grossly deficient, and the head poorly formed and the angle of head and neck undeveloped, but the soft tissues are so contracted, or rather of such inadequate length, that reduction can be obtained even by open operation only at the expense of too great a pressure stress between the head and the acetabulum. The inevitable result is a stiff hip. If such a dislocation is unilateral it may be quite reasonable to accept this stiffness as preferable to a dislocation with its attendant and progressive instability. But if the dislocation is bilateral it is quite unjustifiable to risk two stiff hips. In fact a great authority has gone so far as to say that in a bilateral dislocation of this type, in a boy, the dysplasia is likely to be so great that it is better to leave the hips alone and to deal with any disability at a later stage.

It would perhaps be as well for me to emphasise that in all cases, after open reduction, the hip should be placed, and the limb immobilised, nearly in the anatomical position; then if stiffness supervenes, an important part off unction will be preserved. If on the other hand stiffness occurs with the leg fully abducted, a very different and difficult problem is created.

Student—Have you had experience of this stiffness?

Mentor—In my own patients, no; but on a few occasions I have had to deal with it in others and they made a very marked impression on me.

Student—What procedure did you adopt?

Mentor—I did nothing—just that. There is something about these children apart from the physical which introduces an unknown factor. They are very often an only child, fair-haired and of pale " fragile " complexion. It is as if they react badly to stress. Active treatment like manipulation and passive movements and so on appear to provoke a greater reaction. If they are left to themselves and reassured and provided with some amusing or entertaining diversion, movement gradually returns and a reasonable functional position is achieved. It takes a long time, perhaps as long as two years, and great patience is necessary. While we are on this point perhaps I might give you a warning against open operation immediately after some months of immobilisation. The condition of the soft tissues at such a time will cause them to react unfavourably to the trauma of operation. Also there is a definite limit to the period over which traction should be continued if open operation is contemplated. This is because osteoporosis occurs in the head and neck, and cortical bone seems to turn into eggshell.

Student—What place do you give to rotation osteotomy to counteract anteversion?

Mentor—I am far from being convinced of the value of rotation osteotomy. I believe that there is no place at all for it in the early stage: if the hip is properly reduced, natural remodelling occurs with a return of normal activity. Fortunately Nature is so determined and bone growth is so responsive that the effects of a surgeon's interference soon disappear, " and like this insubstantial pageant faded leaves not a wrack behind,"—or at least only a plate or two and some screws. " De-rotation " may be useful at a later stage, but of more importance is the production or provision of a varus angle in place of the valgus which usually accompanies the type of dislocation we are imagining.

Perhaps I ought to say a word about anteversion. It may be a mistake to imagine that the head and neck pivot on the shaft, and that anteversion will thus cause the head to protrude forwards. Rather must the shaft and the neck pivot on the head. If this is carefully centred in the acetabulum, and kept so for a sufficient time, anteversion cannot cause it to dislocate. On the other hand it is clear that valgus of the neck (in relation to the shaft) can easily cause the head to dislocate, because the head is sliding up the very part of the acetabulum which is deficient. If by osteotomy the valgus is transformed into a varus angle, the thrust of the head and neck into the acetabulum is made normal. Anteversion may be partly or wholly the result of persistent dislocation. In a girl suffering from Still's disease, whose hip had dislocated at the age of five years, anteversion developed to such an extent that at ten years, when open operation was performed, the anteversion had reached 65 degrees (Figs. 18 and 19).

Student—These points need consideration. You keep referring to the head being " centred " in the acetabulum: how can you know this is so in very young patients before the head has appeared radiographically?

Mentor—Well, you have your hands and also you can see in the radiographs that the neck is pointing in the right direction.

Student—I see. And now to return to the question of persistent or recurrent dislocation. Are there not in addition cases in which the neck-shaft angle is good, and yet the hip persistently dislocates even after successful reductions?

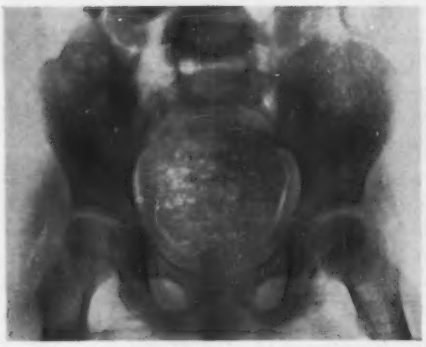

FIG. 18

FIG. 19

Figure 18—A girl aged five years suffering from Still's disease. January 1949. Right hip dislocated during this year. Figure 19—The same patient as Figure 18. November 1954. Right hip dislocated five years previously. Open operation was performed shortly after this radiograph and anteversion of 65 degrees was shown to have developed.

Mentor—Of course there are. Obviously, if the upper part of the acetabulum is deficient and remains so, the head will dislocate when the anatomical position of the leg is reached, and especially when the child begins to walk. And there are cases of subluxation which unfortunately, though naturally, tend to be discovered only relatively late. These are the patients for whom acetabuloplasty is reasonable and useful. Two types of operation have attracted me. First was the Fairbank " shelf " operation. I have studied some results of this in patients on whom it was performed twenty-odd years ago. Two or three years after operation, as cinematograph records show, many were very satisfactory. It has so far been possible to trace only six of these but they have given me much food for thought. It seems that we operated on patients whom we would certainly leave alone today (Figs. 20 to 24). I wonder also whether, after acetabuloplasty of a severely abnormal hip, anything like a normal joint is ever maintained into adult life. It is true that a proportion even of those ultimately poor radiographically were functionally much better than if they had been left alone. Consider, for instance, the girl whose hips are here illustrated (Figs. 25 to 28). She has lived a normal life, and now twenty years after operation she has no pain, she does all her work and can walk " any distance." But for practical purposes might she not have had an arthrodesis of her hip some fifteen years ago? I think that if acetabuloplasty is to be really useful it has to be performed early—except possibly for a subluxation, and then, a shelf just brought comfortably over the unreduced head would be reasonably worth while.

The second type of acetabuloplasty is known as a " Colonna " and has relatively lately come into prominence. In certain instances it has given extremely good results. Its attraction lies in the fact that it brings the head, and therefore the shaft of the femur, nearer the mid-line of the body and so into direct thrust of body weight. The late effects are not yet known. The indications for acetabuloplasty and which type to perform are still being clarified. Broadly speaking it seems to me that up to about five years of age the Colonna method may well be preferable, and after that the Fairbank type. But we may be wrong to expect too much of these operations and we must recognise that there is a limit to the age at which this sort of interference will do good. I would for instance be chary of making fair promises about a child over six years of age with bilateral dislocation. Moreover it may not be unreasonable to perform an osteotomy at a relatively early age if the disfigurement is considerable and the gait grotesque. I have done so at twelve years and have been completely justified by the late result (Figs. 29 to 33).

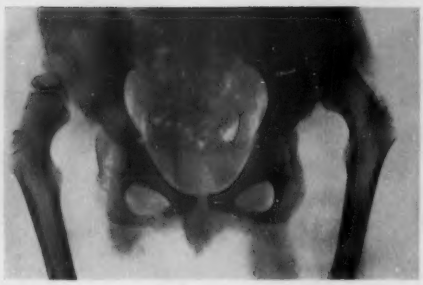

FIG. 20

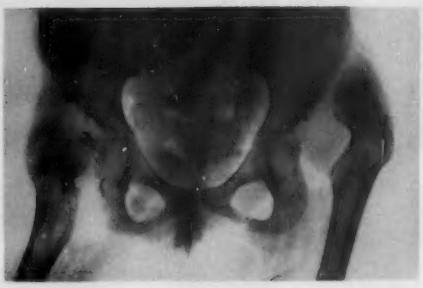

FIG. 21

Figure 20—A girl aged seven years. Untreated bilateral congenital dislocation. (The late Professor McMurray's patient.) Figure 21—The same patient as Figure 20. One year after closed reduction of right hip. Left hip could not be reduced and a shelf operation was performed.

FIG. 22

FIG. 23

Figure 22—The same patient as Figures 20 and 21. Six months after shelf operation on left hip. Figure 23—The same patient one year after shelf operation on left; two years after closed reduction of right. Both hips now dislocated.

FIG. 24

The same patient fifteen years later. No pain but marked limp.

FIG. 25

FIG. 26

Figure 25—The right hip of a girl aged fourteen years, showing untreated dislocation. The left hip was normal.
Figure 26—The same patient as Figure 25. One year later. The right hip has been reduced by prolonged traction and a shelf operation performed.

FIG. 27

FIG. 28

Figure 27—The same patient nine months after shelf operation. Figure 28—The same patient twenty years after shelf operation. She has no pain, does all her own work and can walk " any distance."

FIG. 29

FIG. 30

Figure 29—A girl aged nine years. Untreated. 1930. Figure 30—The same patient. Closed reduction of dislocation.

FIG. 31

FIG. 32

Figure 31—The same patient two years after " successful " reduction. Redislocation. Lorenz osteotomy performed. Figure 32—The same patient six months after osteotomy.

FIG. 33

Same patient twenty-two years after osteotomy. She walks with only a slight limp, does a full day's work as a shop assistant and keeps house for her husband and three children. Up to five years ago would frequently do a twenty-mile walk, but latterly her walking distance has diminished somewhat.

FIG. 34

FIG. 35

FIG. 36

Figure 34—A woman aged thirty-eight years. Right hip very painful. Figure 35—The same case one year after formation of acetabulum and insertion of acrylic prosthesis. Figure 36—Condition two and a half years after operation. The hip is stiff and painful.

FIG. 37

FIG. 38

Figure 37—A woman aged forty-one years. Radiograph two years after Lorenz osteotomy on the right side. Figure 38—The same patient as Figure 37. Radiograph twenty-four years after osteotomy. Has had occasional " rheumatic " pain but never sought treatment because she could walk " any distance " using a stick.

Student—Would you say something about the necessity and the form of treatment for the late effects of congenital dislocation of the hip?

Mentor—You mean about thirty-five to forty years of age? I ought first to say that in certain instances no treatment at all is required. Not infrequently one sees men and women with congenital dislocation of the hip, unilateral or bilateral, who are not complaining of any disability. Quite recently a woman with gross bilateral dislocation attended hospital with one of her children who had sustained some minor injury. The woman was forty-five years old, with a family of four children, and in addition to looking after them she had augmented her husband's income by going out to work. I mention this because the fact puts a limit on the amount and extent of surgery which is justifiable in infancy.

But not all patients escape trouble. Sometimes the dislocated hip itself becomes arthritic and painful and disabling. Many authorities have issued a warning that surgery should be undertaken only for such disability and not merely to relieve limp. Sometimes in cases of unilateral dislocation the affected hip is not complained of, but arthritis has developed in the opposite hip, which originally seemed sound. This is because the instability of the affected hip has thrown extra stress, with resultant excessive wear and tear on its fellow.

Let us first consider this latter type. Reluctant as one is to operate on a part of the body not giving rise to complaint, I have found that stabilisation of the dislocated hip relieves the osteoarthritic hip on the other side to such an extent as to avoid any further treatment. Stabilisation in such a case should be obtained by osteotomy because it would be a mistake to lose all movement by performing an arthrodesis lest the osteoarthritic hip should deteriorate in spite of the relief that has been afforded. I do not advocate arthroplasty with an acrylic prosthesis because, although extremely interesting and quite impressive radiographic appearances can be obtained, the function generally is not good (Figs. 34 to 36).

We have still to consider the management of bilateral dislocation at this middle age. It may be reasonable to perform an arthrodesis on one side, but it must be remembered that this may be difficult because of the poor pelvic bone.

Student—What do you mean by " poor "?

Mentor—The word is used in the sense of hard, thin, sclerotic bone with little osteogenic capacity. Obviously arthrodesis should not be performed on both hips, and if both are painful one at least should certainly be treated by osteotomy. It may in fact be as well to do this for both hips, and in a patient with two painful hips I personally would prefer to perform first an osteotomy on the more painful side. Some freedom of action with the other would be retained. Osteotomy, though unlovely radiographically, is extremely efficient functionally (Figs. 37 and 38).

It is interesting that osteotomy for painful and severely deformed congenital dislocation of the hip in middle life was the practice at the beginning of the period we are considering, thirty years ago. Although in this particular respect we seem to be back where we started, it does not mean that our knowledge is no greater.

Student—Could you now, Sir, summarise what you have said?

Mentor—It is this: Be gentle, be cautious, be patient. Remember the objective of treatment, the principles governing that treatment, and remember that in the management of congenital dislocation of the hip, as of other conditions, there is not one way: rather are there many ways from which you must choose the most appropriate to your patient and to the condition as it presents itself in that patient.

THE TREATMENT OF GAPS IN LONG BONES BY CANCELLOUS INSERT GRAFTS

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The purpose of this paper is to describe a technique for bridging gaps in long bones by the use of solid blocks of cancellous graft combined with internal fixation. This method has proved successful in twenty-seven consecutive cases operated on during the last twelve years. In six of these there had been previous attempts at grafting by other methods and in three cases there had been two such attempts. In two cases operation was followed by a recrudescence of local infection but this did not prevent union. Figure 10 illustrates the kind of problem to which I refer—a gunshot wound sustained in 1944 resulting in loss of bone and a gap of one and a half inches. The original wound had been infected and two previous attempts at restoring union by tibial cortical grafts had failed.

It is common experience that in problems of this kind the classical methods of cortical grafting are not likely to succeed, although they may do so when the ends of the bone can be brought together after freshening. In these circumstances it is usually observed that union occurs between the ends of the host bone remote from the graft, the graft acting mainly as a method of internal fixation. Approximation of the freshened bone ends is not always feasible however (for example, when one of the forearm bones is ununited and the other is intact) and in such circumstances the resultant gap must be accepted and bridged. This problem arises most frequently in non-union of the radius or ulna, but it may also occur in the tibia, femur or even the humerus when apposition of the freshened bone ends would entail too much shortening. It also applies to the bones of the hand, and especially the thumb, in which preservation of length is essential to function.

Before describing the technique that has been used in these cases it is appropriate to consider some of the fundamental principles involved in bone grafting, since they have considerable practical significance.

THEORETICAL CONSIDERATIONS

There has been a great deal of experimental work on the transplantation of bone, originating with the classical experiments of John Hunter and including such famous names as Macewen, Hey Groves, Gallie and Phemister, to mention only a few. In recent years the introduction of the " bone bank " has stimulated fundamental research and revived the old controversy as to whether the cellular elements of a bone graft can ever survive and make a significant contribution to osteogenesis. The experimental evidence on this point is still conflicting. It is established that osteoblasts can survive and proliferate in tissue culture experiments, even after slow freezing (Ray *et al.* 1954), but opinion is still divided as to whether they can survive when transplanted in the body. One hypothesis, for which a vast amount of experimental evidence has been accumulated (Rohde 1925, Campbell *et al.* 1953, Mosiman 1951, Ray *et al.* 1952, Willis 1936) holds that cellular survival can occur provided the graft is autogenous and its texture such as to facilitate rapid diffusion of tissue fluids and early vascularisation (Abbott *et al.* 1947, Ham 1952, Ham and Gordon 1952). These architectural conditions exist only in cancellous grafts. The opponents of this hypothesis hold that any living cells found in such a graft after the first few weeks are the result not of survival but of invasion from the host tissues (Lacroix 1951, Siffert 1955). When the graft is transplanted into an osseous defect such invasion occurs rapidly from the cut ends of the host bone (Siffert 1955) and it is difficult to determine in single plane sections whether a group of living cells in the transplant is the advance guard of such an invasion or a pocket of survivors.

Furthermore, invasion from host sources is the only mechanism which can explain the transformation of dead grafts into living bone (Gallie and Robertson 1919, Phemister 1914, Kiehn *et al.* 1952, Lloyd-Roberts 1952, Wilson 1951). The survival of grafts transplanted into non-osseous tissue such as muscle or the anterior chamber of the eye—as in the experiments of Urist and McLean—might at first glance be supposed to prove cellular survival. But it is not quite so simple because dead transplants have also become repopulated with living osteoblasts in such circumstances (Urist and McLean 1952). It is therefore clear that another mechanism of osteogenesis exists, quite apart from either cellular survival or invasion from host bone. In this third process, which is known as " osteogenesis by induction " (Horwitz 1951, Lacroix 1947), the transplant is invaded by connective tissue cells possessing the pluripotential characteristics of primitive mesenchyme. These immigrant cells are then influenced to undergo differentiation into osteoblasts, but the precise mechanism whereby the transplant exerts this influence is unknown. There are, however, certain circumstances in which the osteogenetic stimulus may become exhausted. The pluripotential mesenchyme then differentiates into fibrous tissue which acts as a mechanical barrier to new bone formation and eventually leads to non-union. In recent important animal experiments Siffert (1955) has observed that this type of reversion is liable to occur: 1) if there is undue movement between the fragments of the host bone and the graft; and 2) if there is delay in the complete impregnation of the graft and graft bed with pre-osseous tissue (*e.g.*, in wide defects). Both observations are of immense practical significance, the first because it stresses the need for absolute fixation, and the second because it emphasises the importance of selecting graft material possessing those physical and architectural characteristics conducive to rapid impregnation. Cancellous iliac graft is ideal in this respect. It offers to the invading osteogenetic tissue a wide flat plain with modern roads and communications. In comparison, cortical bone is an impenetrable forest.

In reaffirming this, Siffert (1955) has added two important new observations. The first is that the larger the absolute surface area of accessible trabeculae in relation to the total volume of the graft, the more avenues there will be for directing the appositional growth of invading osteogenetic tissue, whether this be derived from the host bone, from the surrounding muscle bed or from the proliferation of surviving osteoblasts. These physical conditions are most perfectly attained when the cancellous graft is in the form of a solid block rather than when it is in the form of chips, shaves or ribbons. His second observation is that the liberation of osseo-protein from the cut surfaces of grafts causes a fibroblastic reaction in the invading pre-osseous tissue. The area of cut surface in relation to total volume is minimal in the case of a solid block transplant and this is therefore less likely to induce a fibroblastic reaction than shaves or ribbons.

One interesting practical point has emerged from the experimental work. When young growing animals are used, the osteogenetic response is always much greater than in comparable experiments with adult animals. The inference from this is that a procedure that succeeds in children will not necessarily succeed in adults.

APPLICATION IN PRACTICE

How do these theoretical considerations affect the practical problem of bridging a gap in the radius? First, in the choice of graft material. It is clear both from the clinical and experimental evidence that fresh autogenous cancellous bone is far superior to any other form of transplant (Abbott *et al.* 1947, Dick 1946, Higgs 1946, Watson-Jones 1955). It is the only type of graft in which there is any evidence of cellular survival; it provides the optimum conditions for rapid permeation by invading osteoid from the host bone, and it has the greatest power of stimulating osteogenesis by induction from the surrounding muscle bed. It has further been shown that, for physical reasons, solid blocks give more rapid union and less fibroblastic reaction than chips or ribbons.

The second principle is that the host tissues into which the transplant is made must have a good blood supply, since osteogenesis—whether by survival, invasion or induction—can occur only if the graft is in intimate contact with an active circulation. All sclerotic bone ends must therefore be removed, even though this increases the gap. These sclerotic ends are not only devoid of blood supply but consist largely of fibrous tissue quite incapable of providing an outflow of osteogenetic tissue into the graft. This outflow from the host bone is regarded by Siffert as the most important single source of osteogenesis, so the surgeon must not be afraid of cutting back to normal healthy vascular bone. The gap may look formidable at operation but a two-inch defect between healthy fragments is more likely to be bridged than a one-inch gap between fibrotic fragments.

FIG. 1

Exposure of the ilium. With the patient in the " kidney " position the muscles fall away and the ilium stands up like a ship in dry dock.

The muscle bed must also be freshened by removal of avascular fibrous tissue because osteogenesis by induction can occur if a cancellous surface is presented to a vascular muscle bed, as in the experimental transplants of Urist and McLean.

The experimental evidence also re-emphasises the importance of fixation. It is a first principle of grafting, whether of bones or roses, that there must be intimate and undisturbed contact between graft and host until union is achieved. The easiest and most certain method of doing this (in the case of bones) is by internal fixation with a metal plate, and there is no evidence that this inhibits osteogenesis. A cortical graft screwed to both fragments is a more complicated and somewhat less efficient method of securing internal fixation and it has little osteogenetic value except in children.

The intimacy of contact between host bone and graft is obtained by very accurate shaping and fitting of the graft. This precision is easily achieved with iliac bone, which is rigid enough to be accurately shaped, yet porous enough to be highly osteogenetic.

THE OPERATION

The steps of the operation are illustrated in Figures 1 to 9. The graft is obtained from the antero-lateral aspect of the ilium. In obese subjects, or for cutting large grafts, the exposure

FIG. 2
Removing the cortical "lid." This can either be replaced or used as an "onlay" to reinforce the "insert."

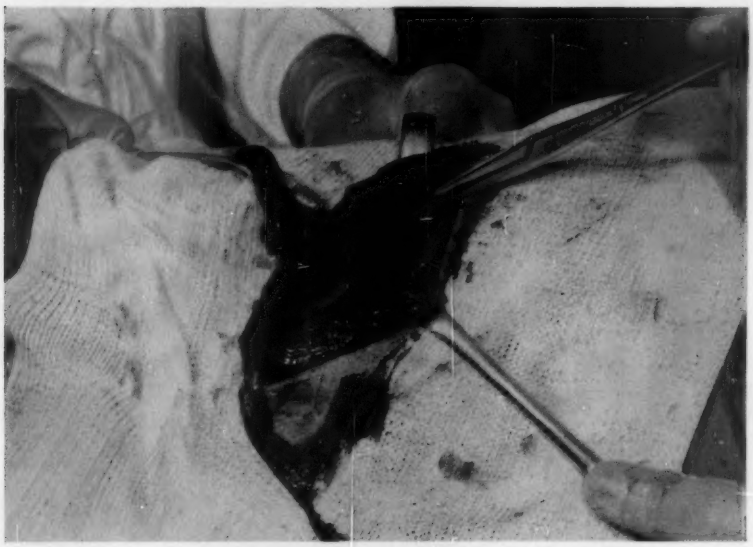

FIG. 3
Reflecting the thin outer cortical plate.

FIG. 4
Removing the graft. In this case the medial cortical plate has been retained.

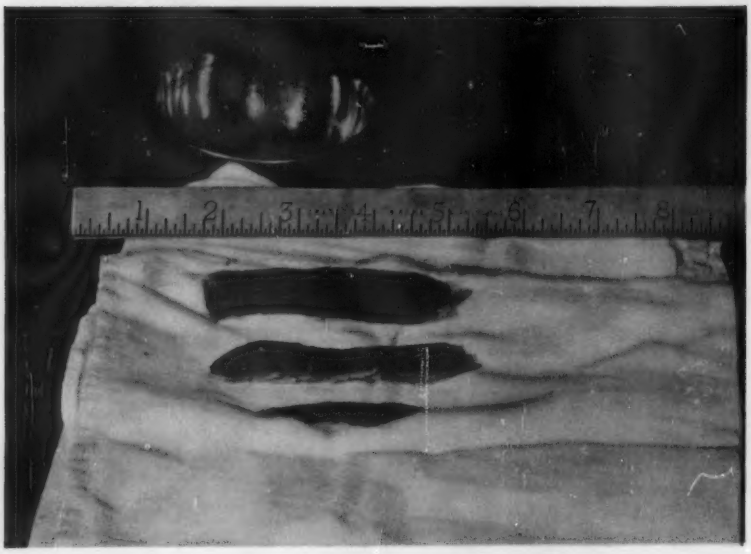

FIG. 5
In a normal subject a solid block of cancellous bone can be obtained three inches long, three-quarters inch wide and half an inch thick. The " lid " is also shown.

FIG. 6
Pseudarthrosis of radius with half-inch gap. Ulna intact. (See Fig. 20.) Trimming back to healthy bone increases the gap to one and three-eighths inches.

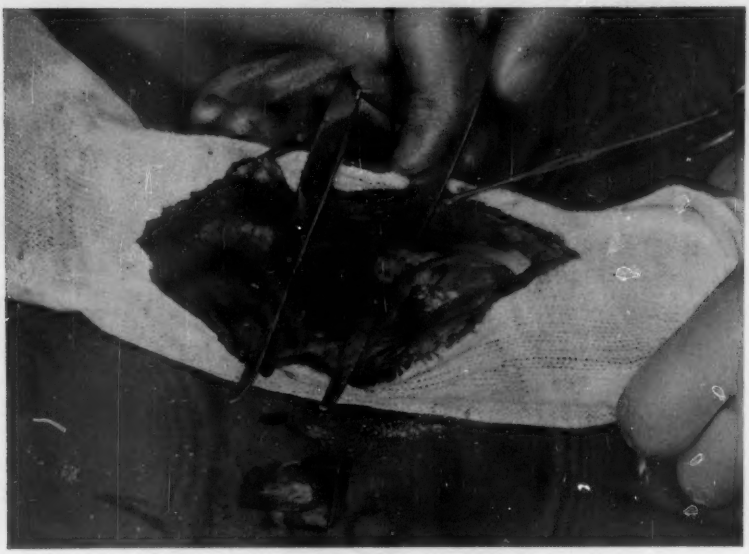

FIG. 7
The pseudarthrosis and sclerotic bone ends have been removed.

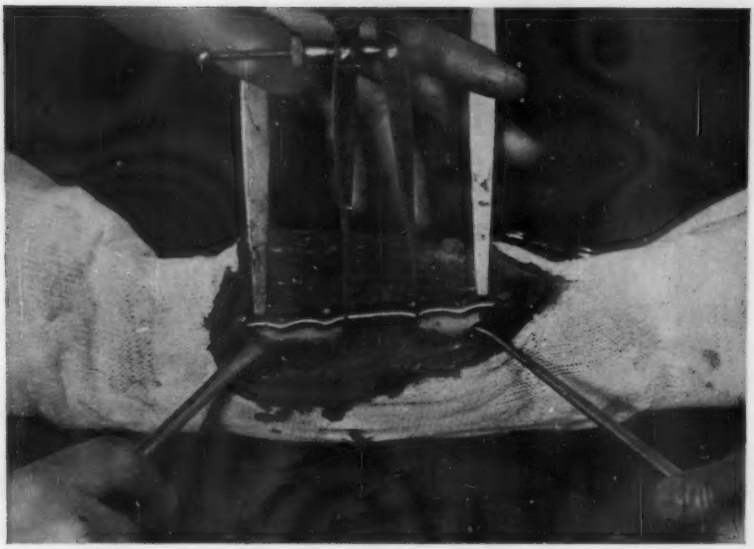

FIG. 8
The fragments have been plated. The gap is being accurately measured.

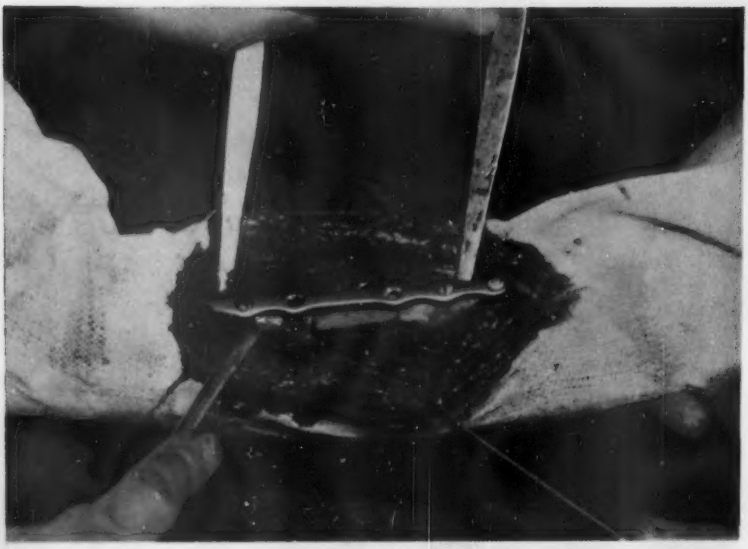

FIG. 9
The graft has been trimmed to the exact length and inserted into the gap. It must be a tight, accurate fit. (The results of this operation are shown in Figures 22 and 23.)

is facilitated if the patient is in the " kidney " position. The superior cortical plate (the " lid ") is removed to expose cancellous bone. It may either be replaced or used as an additional " onlay " to reinforce the " insert." The thin, wafer-like cortical plates on the medial and lateral surfaces of the ilium are reflected and the graft is then cut about half an inch longer than the estimated required length. In all these procedures very thin-bladed osteotomes are necessary—the stone mason's weapons so frequently featured in instrument catalogues are quite useless. In a normal subject a solid block can be obtained up to three inches long, three-quarters of an inch deep and half an inch thick. In large grafts one of the cortical plates is usually retained for strengthening purposes.

FIG. 10 FIG. 11 FIG. 12 FIG. 13 FIG. 14

Case 1. Figure 10—Condition two years after gunshot wound. Two cortical grafts had already failed. Figure 11—Cancellous insert graft. Gap one and a half inches. Figure 12—At four months. Union at the proximal end (normal bone) has been rapid. Union at the distal end (sclerotic bone) is tenuous. Plaster discarded. Figure 13—At nine months. Union firm throughout, but the sclerosis of the distal fragment is still obvious. Figure 14—At eight years. Note re-formation of medullary canal and cortices.

In preparing the fracture site the importance of cutting back to healthy bone has already been emphasised. The fragments are securely plated, the gap is measured with inside calipers and the graft trimmed so that it is a tight fit. The consistency of the graft is such that it can be shaped very accurately with a strong scalpel. If the " lid " is used as an onlay it is tied on with stout catgut. In some cases the internal fixation is so secure that external fixation is unnecessary, but each case must be decided on its merits and no risk must be taken of fixation failing before union is established. In the present series, union has occurred as early as six weeks (Fig. 37) but may take six months in the case of large gaps and avascular tissues (Fig. 12).

SOME ILLUSTRATIVE CASES

Of the twenty-seven fractures in this series fourteen were in the forearm bones, four in the thumb, two in the metacarpals, three in the tibia, three in the femur and one in the humerus. Union was achieved in all cases in an average time of fourteen weeks. Seven cases are presented to illustrate the method.

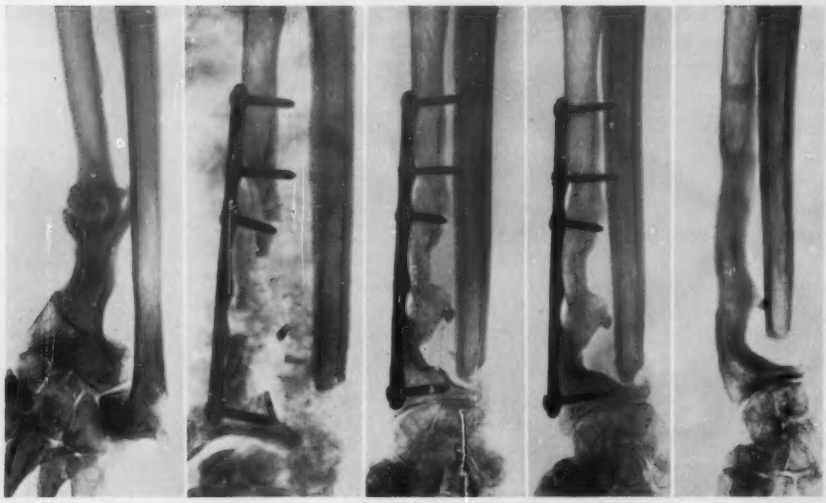

FIG. 15 FIG. 16 FIG. 17 FIG. 18 FIG. 19

Case 2. Figure 15—Twenty-nine years after injury. Figure 16—Cancellous insert graft. Gap after resection of sclerotic bone—three inches. A technically poor operation: the bone has been over-trimmed and badly plated. The graft has been badly cut and does not fit. Figure 17—At six months. Figure 18—At one year. Figure 19—At seven years. Note re-formation of cortices and medulla. Only an operation based on sound principles could forgive the surgeon for such technical incompetence.

FIG. 20 FIG. 21 FIG. 22 FIG. 23

Case 3. Figure 20—Two and a half years after war wound. This is the case illustrated in Figures 1 to 9. Figure 21—Cancellous insert graft. Gap after resection of sclerotic bone ends—one and three-eighths inches. The " lid " has also been used as an onlay. Figure 22—At four months. Union. Figure 23—At fourteen months.

Case 1 (Figs. 10 to 14)—Non-union of the radius with loss of bone resulting from a war wound in 1943. The original wound had been infected and two previous cortical grafts had failed. After a cancellous insert graft, union was rapid at the proximal end, which was vascular bone, but delayed for six months at the distal end, which was somewhat sclerotic.

Case 2 (Figs. 15 to 19)—This pseudarthrosis of the radius resulted from a war wound in 1917. The operation, one of the earliest in the series, was technically poor and deserved to end in failure. The fact that it succeeded is a tribute to the method since only a basically sound procedure could triumph over such technical ineptitude. In contrast there were several cases in which cortical grafts had failed in spite of impeccable technique.

Case 3 (Figs. 20 to 23)—Another war wound (Korea) seen two years after injury, with established non-union of the radius. The fragments were very sclerotic and there was a gap

FIG. 24 FIG. 25 FIG. 26 FIG. 27

Case 4. Figure 24—Pseudarthrosis of ulna after two unsuccessful attempts at cortical grafting. Figure 25—Cancellous insert graft. Gap one and a half inches. Figure 26—At five months. Union. Figure 27—At ten months.

of half an inch which increased to one and three-eighths inches after resection of sclerotic bone. This is the case depicted in Figures 1 to 9 illustrating the operation. Union was sound in four months and recovery of function was complete in six months.

Case 4 (Figs. 24 to 27)—In this pseudarthrosis of the ulna two previous cortical grafts had failed. Both had been performed elsewhere, with impeccable technique, by a surgeon of exceptional virtuosity. A cancellous insert graft was performed two and a half years after the original fracture. The gap in this case was one and a half inches. Union was sound in five months. This patient was away from work altogether for just over three years.

Case 5 (Figs. 28 to 31)—Non-union of the ulna after eight months in plaster. The radius, which was also fractured, has united. The condition was practically identical with that of Case 4 at the time of the first cortical graft. After resection of sclerotic bone there was a gap

FIG. 28 FIG. 29 FIG. 30 FIG. 31

Case 5. Figure 28—Non-union of ulna after eight months in plaster. Radius united. Figure 29—Cancellous insert graft. Gap one inch. No external fixation was used. Figure 30—Union in ten weeks. Figure 31—At eight months. Note how the graft has thickened.

FIG. 32 FIG. 33 FIG. 34 FIG. 35

Case 6. Figure 32—Two-inch gap in tibia following infected fracture with massive sequestration. Figure 33—Twin cancellous inserts. At three months there was evidence of infection and the plate had to be removed. Figure 34—At seven months. There is still a discharging sinus and a small sequestrum has been removed. Nonetheless the grafts have survived and union has occurred. Figure 35—At nine months. Sinus healed. Firm consolidation in spite of infection.

of one inch. No external fixation was used in this case and union was sound in ten weeks. This patient was back at work as a miner four months after operation and a year after injury.

Case 6 (Figs. 32 to 35)—A two-inch gap in the tibia resulting from an infected fracture in which there had been massive sequestration. The skin had broken down in several places and a cross-leg flap had been necessary. Twin cancellous inserts were used, but in three months it became obvious that there had been a recrudescence of local infection and the plate had to be removed. In spite of this the grafts survived and union was present in six months. Cortical grafts in similar circumstances would inevitably have sequestrated. In the tibia and other large bones twin inserts are usually necessary.

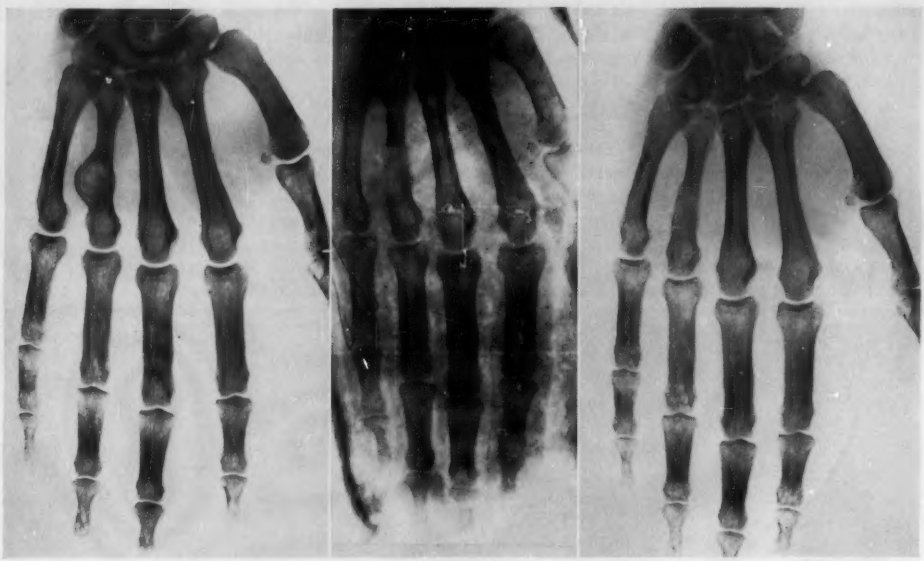

FIG. 36 FIG. 37 FIG. 38

Case 7. Figure 36—Enchondroma of metacarpal. Figure 37—Resection and insert graft (five-eighths inch). Figure 38—At six weeks. Union.

Case 7 (Figs. 36 to 38)—This case illustrates the use of the technique in the bones of the hand. It shows how rapidly incorporation can occur. Sometimes the graft and both fragments are transfixed on a central intramedullary wire.

SUMMARY

1. A method of bridging gaps in long bones is described, using cancellous insert grafts supplemented by internal fixation with a metal plate.

2. The experimental work on transplantation of bone is reviewed in so far as it affects the practical problem of bone grafting.

3. Twenty-seven cases have been treated, with no failures. Seven of these are presented as illustrations.

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FATIGUE INFRACTION OF THE MIDDLE OF THE TIBIA IN BALLET DANCERS

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In six years we have encountered an unusual defect of the shin in four ballet dancers and have carried out biopsy in two. A fifth dancer possibly had the same defect before a complete fracture of the tibia from muscle action. The chief symptom has been localised pain on leaping. A tender swelling has been found at about the middle of the tibial crest, where radiography has revealed a horizontal fissure with adjacent hyperostosis.

CLINICAL MATERIAL

Case 1—Defect of left tibia. Biopsy. A male ballet dancer aged twenty-three began to suffer a stab of pain in the middle of the front of his left shin when landing on this leg from leaps. A few days later while pulling on his socks he chanced upon a small tender hard lump at about the middle of the crest of his tibia. He did not limp, and he paid no attention at first, but the lump became larger and more tender.

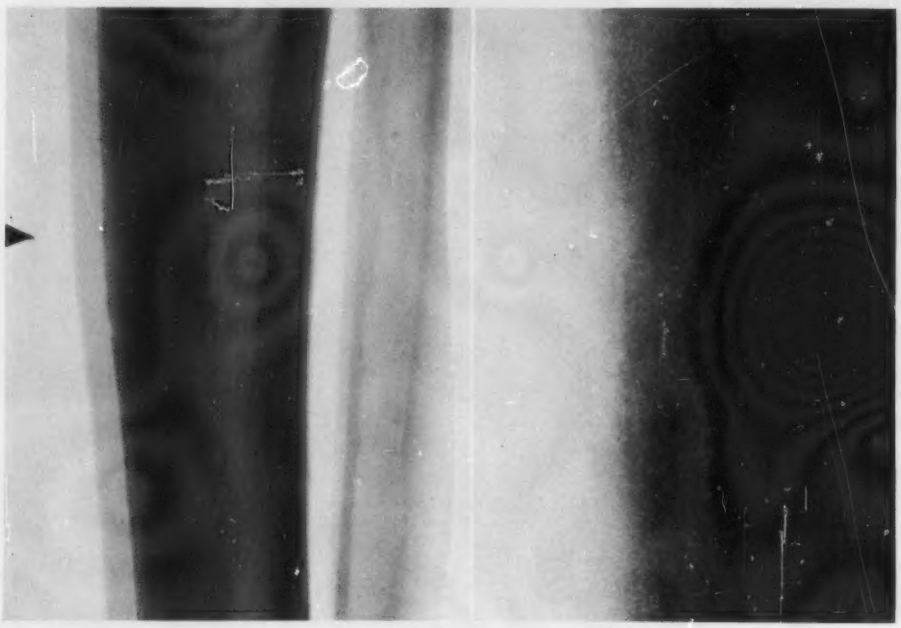

FIG. 1

Case 1—Radiograph a few days after onset of pain. (Right-hand radiograph, ×2.)

Six weeks before he had attended elsewhere for an inversion sprain of the right ankle, which had since recovered almost fully.

On examination, palpable irregularity was confirmed at the site mentioned—but without tenderness on pressure or percussion, and without pain on movement except jumping.

Radiographs (Fig. 1), taken by Dr Campbell Golding, in various projections and with a marker over

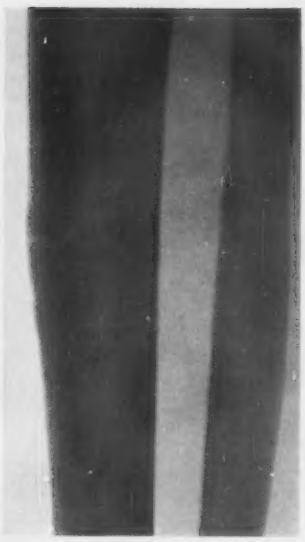

FIG. 2

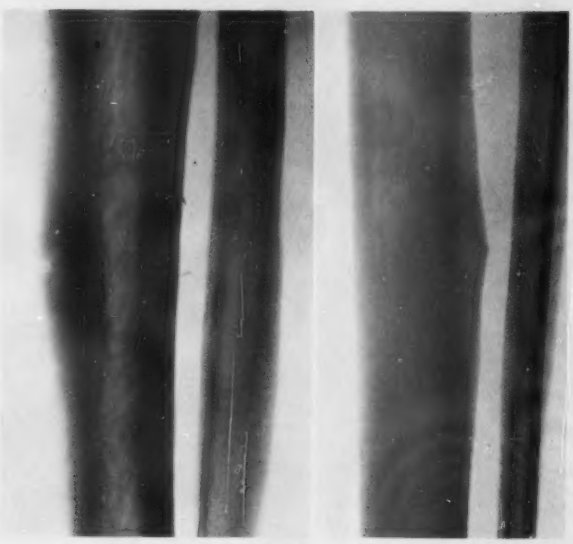

FIG. 3

Case 1—At five months and ten months, the radiographs show progressive hyperostosis of the front of the tibia with a clear intersection.

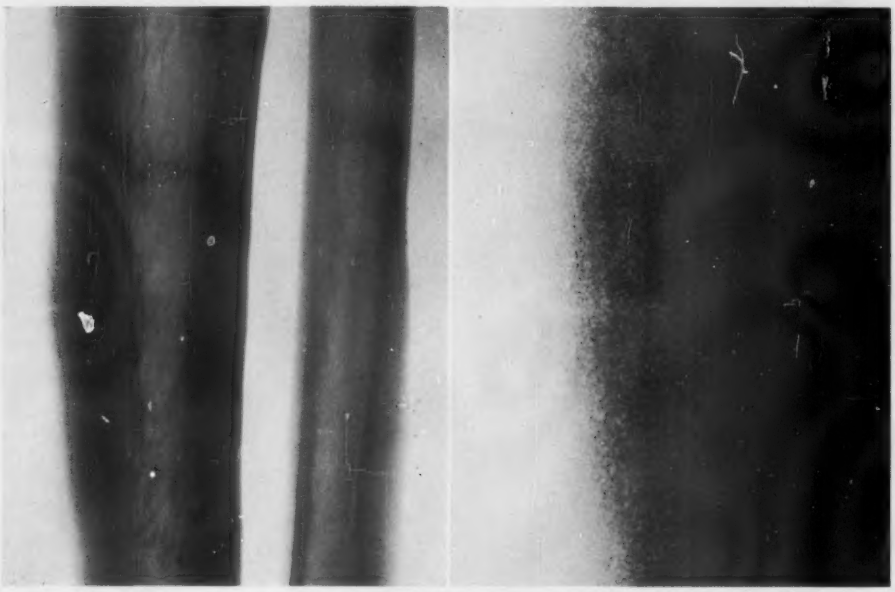

FIG. 4

Case 1—Six years and eight months after the onset, a defect was still evident although symptomless. (Right-hand radiograph, ×4.)

the slight prominence, showed very slight irregularity here with a just perceptible underlying linear rarefaction running into the bone for 3 millimetres. The tibia was not measurably thickened. The Wassermann and Kahn reactions were negative.

During the next five months the patient carried on normally, but the pain steadily increased and the hard swelling of the front of the tibia gradually enlarged. It was still not tender, but the overlying skin, otherwise normal, was slightly hot. Pain was provoked by walking or dancing and relieved by rest. The radiographs now (Fig. 2), and five months later still (Fig. 3), showed a progressive increase in the fissure and the hyperostosis.

Biopsy—Fifteen months after the onset a block of bone was removed. This included the fissure, into which the periosteum seemed to sink. The rest of the hyperostosis was removed; it appeared whiter than the surrounding bone and was very hard, tending to flake beneath the osteotome.

Histology (Dr H. A. Sissons)—The cleft contained only a little fibrinous débris and fibrous tissue, without callus. The adjacent bone was normal except for some enlargement of the central vascular canals of its Haversian systems.

Progress after biopsy—The patient resumed ballet exercises four weeks after operation and full dancing at six weeks. A subsequent fracture of the base of the right fifth metatarsal bone from a supination injury healed normally.

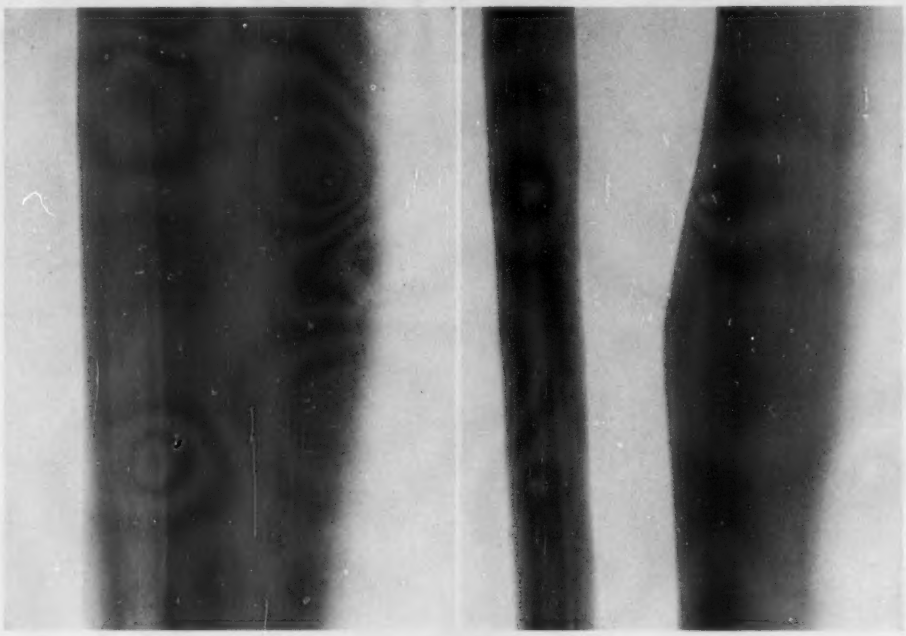

FIG. 5

Case 2—Mid-tibial crest when first radiographed, twenty months after onset; cortical thickening and intersection.

Six years and eight months after the onset (five years and five months after the biopsy) he was still at full work having had no further trouble whatever with the leg, which was normal on examination except for the operation scar. A radiograph still showed the defect, but faintly (Fig. 4). Further blood investigations were normal (serum calcium 9·2 milligrams per cent; serum alkaline phosphatase 5·8 King-Armstrong units) except for a slightly high serum inorganic phosphate content of 4·0 milligrams per cent.

Case 2—Defect of right tibia. (Presumed old fatigue fracture of second right metatarsal bone.) Biopsy. A male ballet dancer aged twenty-six was kicked on the right shin twenty months before, and had a further blow in the same place a few days later. After ten days' rest the pain disappeared and he resumed work. When jumping he began to notice gradually increasing local mid-tibial pain both on taking off and on landing. There was no pain with any other activity. Seven months later

FIG. 6

Case 2—Lateral cortical thickening of second right metatarsal bone, possibly representing old fatigue fracture.

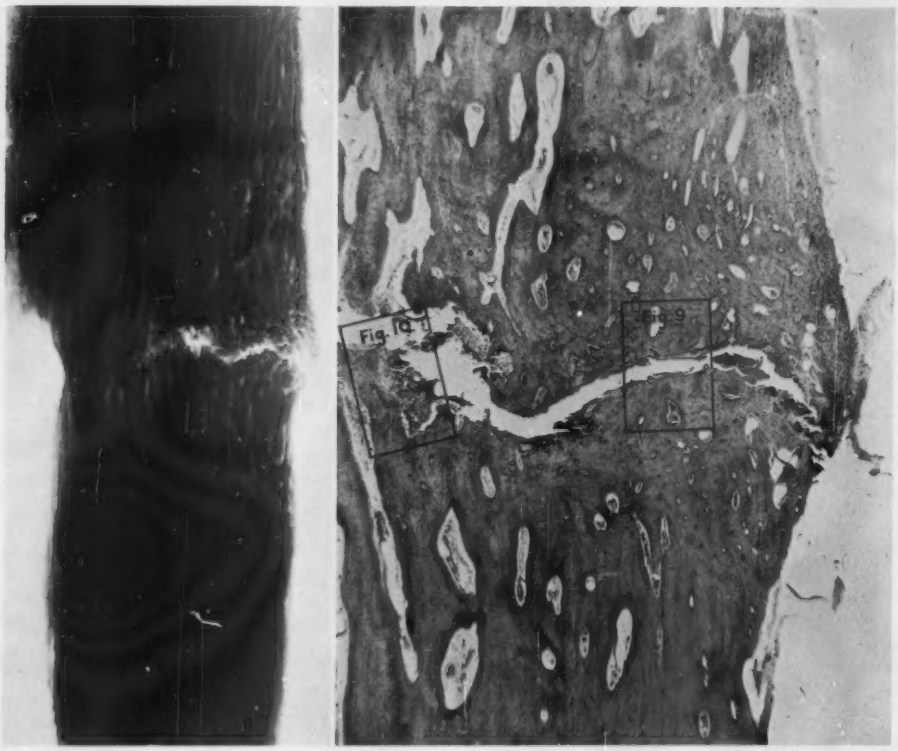

FIG. 7 FIG. 8

Case 2. Figure 7—Radiomicrograph of biopsy specimen including the infraction; the periosteal surface is to the right. (×4.) Figure 8—Low-power photomicrograph of bone adjacent to the fracture defect. The periosteal surface of the bone is to the right. Deep to this, and traversed by the defect, is an area of newly formed bone tissue showing numerous small vascular channels. The rest of the bone in the field is compact cortical bone showing considerable enlargement of the vascular spaces of its Haversian systems. (×16.)

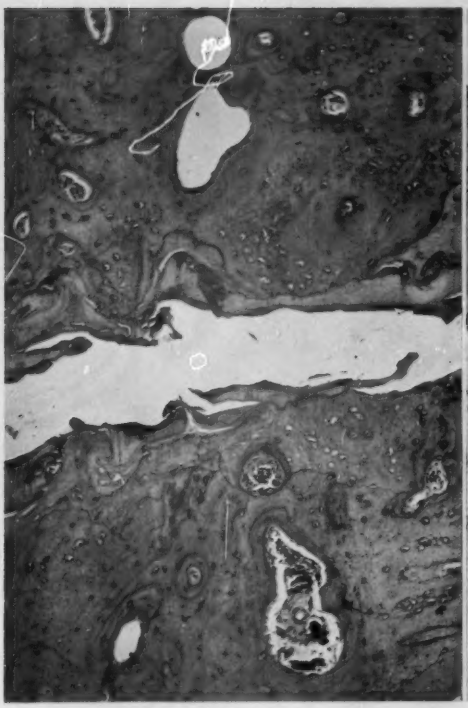

FIG. 9

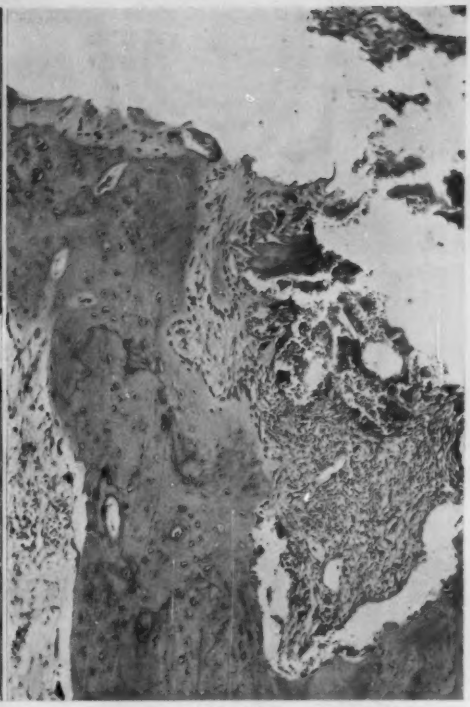

FIG. 10

Case 2. Figure 9—Histological structure of newly formed subperiosteal bone. The surface of the defect consists of normally calcified bone, partly covered by a layer of hyaline fibrinous material. The adjacent bone shows occasional empty lacunae. No excess osteoid tissue is present. (×90.) Figure 10—An area where the surface of the cortical bone adjacent to the defect shows a small area of vascular granulation tissue containing a few necrotic bone fragments. (×90.)

FIG. 11

FIG. 11

An area of the deeper cortical bone adjacent to the defect. The vascular spaces of the Haversian systems are greatly enlarged. The bone immediately adjacent to the defect shows occasional empty lacunae, but is not otherwise abnormal. (×90.) (Dr H. A. Sissons.)

his doctor in Canada drew attention to a hard, tender ridge. He had radiographs taken, which were said to show a tibial fracture, and carried out a local operation. The patient resumed work after two months; but in the last eight months the pain was worse. Five weeks' rest from dancing failed to bring relief. He had no limp.

On examination, the only abnormal physical sign, except the scar of a presumed biopsy, was slight localised tenderness on strong pressure over the middle of the right tibial crest.

Radiographs showed at this point cortical thickening, half-transected by a horizontal crack (Fig. 5). No lesion was found elsewhere in the skeleton, except thickening of the lateral cortex of the second right metatarsal bone (Fig. 6).

Blood investigations were all normal—Hb. 14·8 grammes per 100 millilitres. W.B.C. 6,500 per cubic millimetre (P.4,290, L.2,015, E. 195, B. 0, M. 0). W.R. and Kahn tests negative. E.S.R. 5 millimetres/1 hour (Westergren). Serum calcium 10·6 milligrams per 100 millilitres. Serum inorganic phosphate 3·2 milligrams per 100 millilitres. Serum alkaline phosphatase 6 King-Armstrong units.

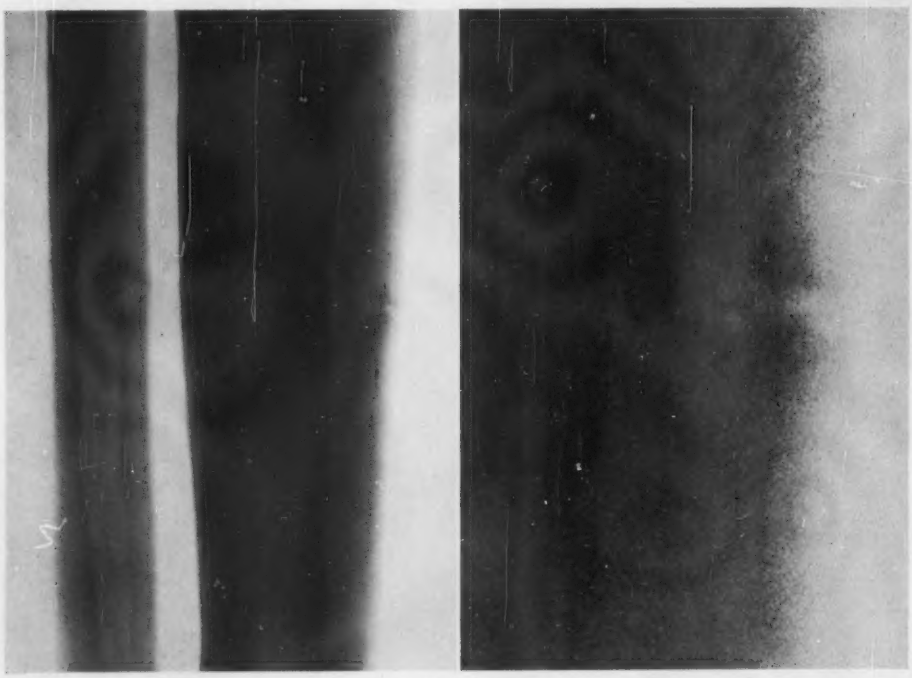

FIG. 12

Case 2—Three years and nine months after the onset, a defect is still visible. (Right-hand radiograph, ×4.)

Biopsy was performed twenty-two months after the onset. The locally thickened and sclerotic area of tibia was removed, together with a minute red horizontal fissure just discernible on the crest.

Histology (Dr H. A. Sissons) (Figs. 7 to 11)—The cleft was occupied by fibrinous débris and a small amount of vascular granulation tissue, without callus. On either side of it superficially there was some subperiosteal new bone, and more deeply the adjacent cortical bone showed rarefaction from enlargement of the Haversian canals, which contributed to the transradiancy of the defect. The remaining bone was normal.

Progress after biopsy—Despite some initial muscle stiffness and the formation of considerable subperiosteal radiating new bone, the patient made good progress and returned to full work. Now forty-five months after the onset (twenty-three months after the biopsy) he still has a little pain on taking off and on landing if he goes hard at a leap, so that he has "cut down on them a little"; nobody notices, and the slight trouble is becoming no worse. No tenderness or swelling has returned. A fresh radiograph (Fig. 12) still shows a small defect; the anterior tibial cortex is thinner than in the opposite leg, but the posterior cortex is thickened.

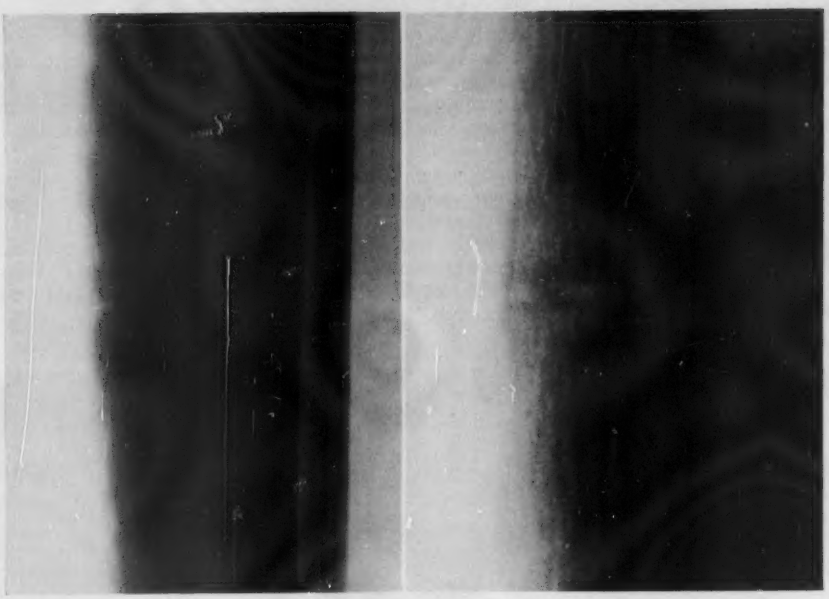

FIG. 13
Case 3—First radiograph, eight to ten weeks after onset of pain. (Right-hand radiograph, ×4.)

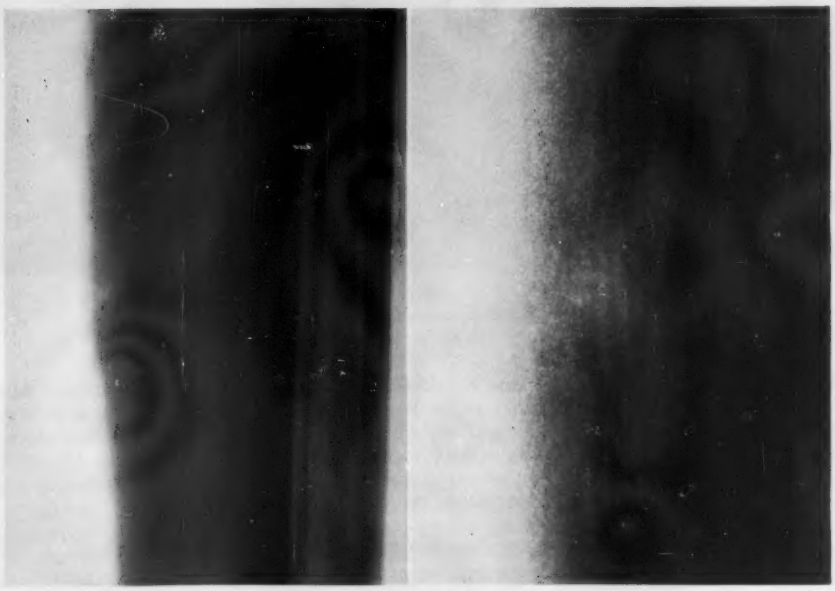

FIG. 14
Case 3—Thirteen weeks later. (Right-hand radiograph, ×4.)

Case 3—Defect of left tibia. More recent case. A male ballet dancer aged twenty-one had noticed, eight to ten weeks before, the gradual onset of diffuse aching in the front of the left shin, especially on forced dorsiflexion or plantar flexion of the ankle, or on negotiating stairs—going up more than down. There was no pain except when weight was taken. For four weeks he had been unable to jump on his left leg, and there was localised tenderness of the middle of the front of the shin.
Examination revealed a tender localised swelling of the middle of the tibial crest and slight tenderness of the corresponding subcutaneous surface. *Radiography* showed a defect in the thickened tibial crest (Fig. 13).

With rest from work the swelling diminished and a groove became palpable across it. Radiographs showed improvement. After only thirteen weeks the intersection appeared to have been replaced by a faint, buried, rounded area of rarefaction slightly reminiscent of osteoid osteoma (Fig. 14). He then resumed work—gradually, intermittently, and with pain on jumping. Now, thirteen months after the onset, he is at full work on tour abroad with the first company, and reports, " My leg is giving me no trouble at all even when I do a lot of jumping, although at times it is slightly painful to touch at the spot where the fracture occurred."

FIG. 15
Case 4—Our only case in a woman. (Right-hand radiograph, ×4.)

Case 4—Defect of the left tibia. Only case in a danseuse. With very prolonged and arduous dancing in the ballet *Carmen*, a French ballerina in her twenties suffered gradually increasing pain at the middle of her shin, which presented a tender mid-tibial swelling. She could not recollect any injury. Radiographs showed a linear defect and some thickening of the left tibial crest (Fig. 15); the right tibia was normal. The pain and swelling increased, and after a year the swelling was excised in France. No further details are obtainable except that she returned to full dancing.

Case 5 is of speculative interest, but is included as a possible example of a serious complication, spontaneous fracture, already reported in fatigue infraction of the tibia (Hartley 1942, 1943). The patient noticed a tender swelling at the middle of the tibial crest; this proved a site of weakness, because a few days later the tibia broke here with muscle action. The initial tender lump may have indicated the condition found in Cases 1 to 4.

Case 5—Complication of presumed defect of left tibia. Acute fracture from muscle action. This ballet dancer aged twenty had suddenly begun to notice left mid-tibial pain whenever he took off in a jump; a week later this complaint had drawn his attention to a slightly tender hard lump half an inch in diameter on his left shin bone. Thereupon radiographs had been taken of the ankle and lower six inches of the tibia; they omitted the site of complaint, and were pronounced normal. There was no limp. About seven days later, just as he leapt, he felt and heard a " colossal crack " in the same place. Sure that he had broken the leg, he landed on the other one and did not try to walk. *Radiographs* showed a comminuted fracture of the middle of the tibia (Fig. 16).

A satisfactory position was maintained in plaster-of-Paris. There was clinical and early radiological evidence of union at seventeen weeks. Partial weight bearing out of plaster was then allowed with crutches and sticks; but twelve days later he stepped smartly out of the way of a bowl

FIG. 16
Case 5—Comminuted fracture of mid-tibia and of fibula from indirect violence after symptoms suggesting the defect here described.

at a rehabilitation centre, felt a crack in the left leg, and was unable to stand on it. Clinical and radiological examination confirmed refracture. With five months' plaster-of-Paris support the fracture united posteriorly; a gap remained in front for a further two months.

Fourteen months after the original fracture there were firm consolidation and full joint movement. He had become a stage manager, but still took part in dancing which, three months later, still caused an occasional ache and an occasional " twinge " at the site of fracture when he took weight on his left leg alone. The girth of the left calf was half an inch less than the right.
Radiographs remained satisfactory.
Pathological investigations gave normal results (serum calcium 9·5 milligrams per 100 millilitres; serum alkaline phosphatase 7–9 King-Armstrong units) except perhaps for a rather high serum inorganic phosphate content (4·1 milligrams per 100 millilitres).

DISCUSSION

THE CHARACTERISTICS OF THE SYNDROME

The patients were young, healthy ballet dancers. Though women greatly outnumber men in the profession, only one of our patients was a woman. This may be fortuitous, but there are several possible reasons why male dancers might be more susceptible. Boys often take up ballet later than girls—as late as fifteen to seventeen years of age in our male patients, who nevertheless were all in top companies. The man's legs have often to carry his partner's weight as well as his own. The man's position on " half-points " is reputed—for what this is worth—to throw more strain on the calf than the stance of the danseuse on her " points." The man's leg is heavier and consequently its inertia is greater. But possibly the most important source of trouble is the man's great and magnificent leaps; it was the take-off in these that gave pain.

The site—The lesion was always at about the middle of the tibial crest. Only one leg was affected; some of the dancers considered it the dominant one.

The symptom common to all the men was gradually increasing pain, confined or almost confined to the take-off in jumps and never present during rest. The character of the danseuse's pain is unknown. No injury was recollected except in Case 2.

The signs—All the patients presented tenderness and a palpable lump, but the pain was always noticed first.

Radiological appearances—The most obvious thing was a horizontal fissure extending into the cortex of the tibial crest, which was itself thickened by subperiosteal new bone with a slightly irregular surface. The fissure lay mainly, but not wholly, in the new bone, and seemed to extend only a short way into the original cortex. Sometimes it showed a finely pencilled outline of increased density. The changes were best seen in a lateral view; but they were also discernible in the antero-posterior view, in which the swelling might be mistaken for part of a well formed tibial crest, a supposition disproved by symmetrical radiography of both legs.

Pathological examinations—Biopsy in two cases revealed no evidence of general disease but a complete absence of callus in the defects.

THE NATURE OF THE DEFECT

The radiological picture of the fissure, with the adjacent hyperostosis, is very reminiscent of some examples of Looser's *umbauzonen* of the osteomalacias, in which a well outlined fissure with pouting mouth may partly penetrate a long bone, often on its concavity. Similar looking defects occur in Paget's disease, but on the convexities (Fig. 17). The term pseudofracture has been applied to all these seeming infractions (incomplete fractures) in pathological bone, but has not yet been accurately defined. The histological appearances in our two cases subjected to biopsy exclude the possibility of general bone disease.

More rarely, the same radiological appearance of fissure and hyperostosis is found in incomplete fatigue fractures of the tibia (Roberts and Vogt 1939, Hartley 1942, 1943). The occupational incidence in our patients, taken with their good health, points insistently to fatigue or stress as the cause of the defect, and we conclude that it is a fatigue fracture—or more precisely a fatigue *infraction*, because it involves only part of the diameter of the bone.

Most fatigue or stress fractures of the tibia happen in the upper third. Here they are almost confined to children and adolescents, and, if incomplete, present the defect posteriorly. Fatigue fractures do however exceptionally happen at the middle of the tibia.

The level of a fatigue fracture in a bone, and the aspect on which it starts, must depend among other things upon the sites of greatest stress—not only the stresses of weight bearing,

but those of movement. A bending force applied to a rod or tube must produce distraction stresses on one aspect and compression stresses on the other. It may be that distraction is more important than compression in starting a fatigue fracture and therefore in determining the orientation of an infraction. Consequently it would be understandable that a ballet dancer, who performs almost acrobatic movements, should crack the bone at a different level and on a different aspect from, say, a boy in a naval training establishment, who always moves at the double.

The failure of these infractions in normal bone to display the abundant callus and ready union of ordinary fatigue fractures may perhaps be attributed to distracting forces that continue if the provocative movement is persisted in. Probably more important is the virtual absence of play between the surfaces of an infraction. Completion of the fracture leads to ready union (Hartley 1942). A parallel may be drawn by the infractions in Paget's disease. Situated on the convexity of a bowed bone, they are subjected to a distracting force, and, being incomplete, they are protected from movement. If the fracture is completed they readily heal with the abundant callus so characteristic of Paget's disease.

FIG. 17

Tibia in Paget's disease showing characteristic infractions on the convexity from bending stress in a curved structure of poor comp › ition.

DIAGNOSIS

What has been said of this lesion, and particularly the disaster of Case 5 (whatever its true nature), confirms the importance of meticulous clinical and radiological examination. In doubtful cases multiple radiographs are needed with a marker at the site of tenderness, and the films should be scrutinised through a magnifying glass. The minute changes could easily be missed by a competent and careful clinician.

The clinical resemblance of osteoid osteoma and the radiological resemblance of infraction in bone diseases, such as osteomalacia and osteogenesis imperfecta, must be borne in mind.

TREATMENT AND PROGNOSIS

Though four patients have improved with rest, with or without biopsy, and have returned successfully to their strenuous occupation without prolonged immobilisation, the cures have not all been complete in the sense that every patient can use his full powers in leaping without a reminder of the lesion and that no evidence of the fissure is to be found in radiographs. A proper assessment will be possible only with the elapse of more time and the study of more cases.

When the diagnosis seems reasonably certain the best treatment may be immobilisation in a long walking plaster incorporating the knee and foot.

When the diagnosis is in doubt it may perhaps be better sometimes to remove a block of bone containing the fissure and fill the gap with an inlay graft.

At present it is impossible to say when work can safely be resumed. To await radiographic restitution may be to postpone dancing indefinitely; some risk may have to be taken. Attention to tenderness and to graduation of work are perhaps the best safeguards.

SUMMARY

1. A defect of the middle of one tibial crest is described in young healthy ballet dancers.
2. The clinical and radiological characteristics have been studied in four cases, and the histological appearances in two.
3. The defect is concluded to be an incomplete fatigue fracture—a fatigue infraction.
4. A fifth, uncertain, case has been added tentatively, as presenting a possible serious complication, namely acute fracture from muscle action.
5. The diagnosis, treatment and prognosis are mentioned, with emphasis on the importance of adequate clinical and radiological examination in cases of obscure mid-tibial pain, tenderness or swelling.

I have to thank Dr E. F. Scowen for referring most of these patients, Dr H. A. Sissons for the histological investigations, and Dr Campbell Golding for some of the radiography; from all these I have had valuable help and advice, as also from Dame Ninette de Valois. I am indebted to Mr D. V. Webb for photomicrography, and to Mr R. J. Whitley and Mr H. G. Berkshire for other photographic work.

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CHONDROMALACIA OF THE PATELLA *

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From the Middlesex Hospital and the Bland Sutton Institute of Pathology

Chondromalacia is a precursor of osteoarthritis. It can, perhaps, be regarded as an early senescence (the changes in the intervertebral discs that begin during the second decade of life are often so described) but much more must be learnt about the chemistry of proteins and polysaccharides before such an idea can be accepted. The articular cartilage of the patella is involved earlier and more constantly than almost any other joint surface. The process often begins during the second decade of life, and by the age of thirty nearly everyone is affected. It is, however, in only a few individuals that the changes cause symptoms, and in fewer still that they progress to osteoarthritis. The late stage, osteoarthritis, has been familiar for centuries. Descriptions of the early phases are comparatively recent, and even though a clinical diagnosis of chondromalacia of the patella can now be made with some precision, the condition has received but scant attention in the literature of British and American countries.

Credit for the first account is generally given to Büdinger (1906, 1908) who described fissures occurring in the articular cartilage of the knee, usually the patella, which he considered to be traumatic in origin. He operated on fifteen patients, removing the affected cartilage in thirteen with results that were, in the main, satisfactory. Ludloff in 1910 published one case in which he excised almost all the cartilage covering the patella, and Axhausen (1919) reported another in which he had removed part of the cartilage six years previously with a good result. In 1925 Läwen reported the results in thirteen operations and also that he had found similar changes in twenty-six necropsies out of forty-four. The term " chondromalacia " seems to have been introduced by Aleman (1928) who found degeneration of the articular cartilage of the patella in one-third of 220 knees on which he had operated, and in twenty this was the only lesion. There were several further reports during the next few years, and in 1936 Øwre published an extensive survey giving the results of a painstaking clinical and pathological investigation, and his findings in thirteen operations. Chaklin (1939) stated that he had removed the articular cartilage of the patella thirty-eight times, but gave no details of the results. In 1940 Karlson reported the results of operations carried out in Aleman's clinic, and also the subsequent behaviour of seventy-one knees not subjected to operation. Wiberg (1941) investigated the etiology with special reference to the congruity of the joint surfaces, and Hirsch (1944) reported certain physico-chemical investigations and forty-one operations; Soto-Hall (1945) treated six soldiers by excision of the patella, and Cave, Rowe and Yee (1945) reported eleven more in whom they had removed the cartilage. In 1948 Gray gave a summary of the morbid anatomy and clinical findings and reported eight patients treated by excision of the patella with good results. The total number of operations reported is more than 200.

INCIDENCE

The first microscopic change in the articular cartilage is absence of the normal metachromasia. The earliest macroscopic change is swelling and softening of the cartilage—often misnamed oedema—which is present in at least five out of six people by the age of thirty. The next stage is fissuring and flaking (Fig. 2), and this can be found in more than

* A part of this paper was read by one of us (P. W.) at the Annual Meeting of the Canadian Orthopaedic Association on June 28, 1955.

half the population by thirty, the incidence increasing steadily thereafter. The figures obtained by Øwre (1936) at post-mortem examination are shown in Table I.

The frequency with which fissuring of the articular surface of the patella is found at operation for removal of a semilunar cartilage is somewhat less than these figures suggest, perhaps because of the restricted view available. The severity of fissuring cannot readily be recorded statistically because estimation is a matter of personal judgment. Øwre did not find at necropsy any example under the age of thirty that he could classify as severe, but there

TABLE I

INCIDENCE OF CHONDROMALACIA OF THE PATELLA IN 124 NECROPSIES (ØWRE)

Age	Number of subjects	" Oedema "	Fissuring
14–19	18	5	—
20–29	32	27	18
30–39	26	24	19
40–59	32	30	29
60–80	16	16	15

were nine between thirty and thirty-nine, and forty over the age of forty. These findings are not in complete accord with clinical experience. The commonest age for symptoms to arise is between twenty and thirty, and we have often found advanced changes at operation on patients of this age and, indeed, in younger people. The discrepancy is probably explained by the natural selection of those attending for clinical examination of the knee.

It is difficult to estimate how often chondromalacia gives rise to symptoms but it is clear that the clinical diagnosis becomes more common as awareness of the condition increases; and it is more often seen at operation when it is deliberately sought. Some idea of the incidence is given by comparing the frequency with which chondromalacia with marked fissuring has been observed at operation with the number of meniscectomies performed during the same period. The figures for the Middlesex Hospital, including chondromalacia encountered during meniscectomy, are shown in Table II.

TABLE II

INCIDENCE OF CHONDROMALACIA OF THE PATELLA AS SEEN AT OPERATION RELATED TO THE NUMBER OF MENISCECTOMIES PERFORMED DURING THE SAME PERIOD (MIDDLESEX HOSPITAL)

	1950	1951	1952	1953	1954	1955 Jan./June	Total
Chondromalacia	4	2	8	5	22	7	48
Meniscectomy	31	24	37	29	32	13	166

NATURAL HISTORY

The natural evolution of chondromalacia cannot readily be followed because the precise condition of the cartilage cannot be estimated with accuracy by clinical examination alone; and when there are symptoms severe enough to call for operation, the affected cartilage (or the patella) is generally removed and its subsequent behaviour cannot be observed.

Nearly all adult knees show some of the pathological changes of chondromalacia, but these are generally restricted to a small part of the patella. Progress, in a large majority, is

FIG. 1

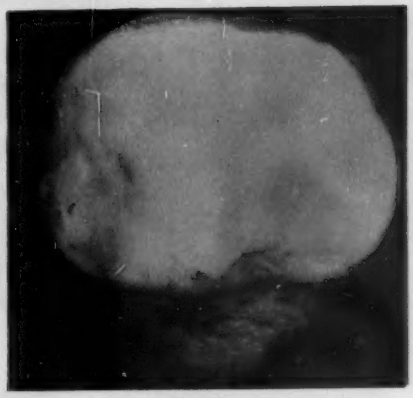

FIG. 2

Figure 1—Erosion of the lower and inner quadrant surrounded by areas of nodular swelling and fissuring, and covered with pannus. As seen at operation May 1955. Figure 2—Erosion of the lower and inner quadrant with fissuring above, and also fissuring of the lower and outer quadrant. Pannus is spreading over the lower border. Case 40.

FIG. 3

FIG. 4

Figure 3—The whole surface of the patella is disintegrating. Patella excised June 1955. Figure 4—Early osteoarthritis. The articular cartilage of the patella and the inner condyle of the femur has been shed. As seen at operation after excision of the patella October 1954.

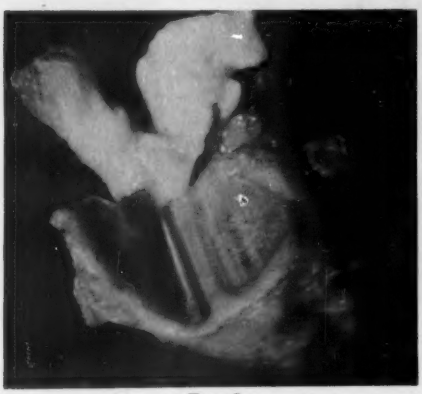

FIG. 5

FIG. 6

Figure 5—Advanced osteoarthritis with a large marginal osteophyte and vertical scoring. Figure 6—Same patient as Figure 5 showing vertical scoring of femur. As seen at operation June 1954.

so slow that there are no symptoms throughout life. In a few the changes, almost from the start, are more widely spread, and in some of these progress is rapid. Occasionally it is very rapid; there is a type in which the entire surface of the patella disintegrates and, when the knee is opened, the synovial fluid is clouded by fragments of articular cartilage. There is at present no sure way of telling by clinical examination, or even by inspecting the articular cartilage, whether progress will be slow or quick, nor is there a constant relation to any feature of the history, such as injury. The determining factor is more likely to be found in the chemical behaviour of the ground substance of the cartilage.

Something is known about the average, or statistical, behaviour of chondromalacia after a clinical diagnosis has been made. Karlson (1940) re-examined seventy-one men who had been diagnosed in Aleman's clinic between one and twenty years previously when they were army conscripts, and whose symptoms did not then warrant operative treatment. Ten had no subjective complaints, forty-five had a little trouble such as stiffness and fatigue after severe exertion, and sixteen had had enough trouble to make them change their work or to cause frequent absence from work. Thirty-five were radiographed and seven showed slight osteoarthritic changes but none had advanced arthritis.

We are fortunate enough to have seen and recorded the presence of fissuring in nineteen knees operated upon for other conditions, the articular cartilage being left intact. Seventeen of these operations were between one and five years ago. In eleven knees only a small area of cartilage was affected; four now have no symptoms, five have a little aching or swelling after exercise and two cannot be traced. The fissuring was of moderate severity in four: one now has a " normal " knee, two have troublesome symptoms and one cannot be traced. Two patients, both operated upon three years ago, then had severe and extensive changes in the cartilage, and now both have clinical and radiological signs of osteoarthritis. These observations suggest that the more widespread the changes the more likely they are to progress.

Chondromalacic changes are quite often seen on the femur at sites corresponding to those on the opposing surface of the patella, but this is by no means invariable. There may be advanced changes in the patella without any visible alteration in the femur, or those in the femur may be more advanced than the patella, and occasionally the femur alone is affected. It seems that a rough patellar surface rubbing on the femur is not the only cause of the latter's degenerating. It may be that, as in the patella, a predisposing factor must be postulated.

Chondromalacia of the patella sometimes progresses to osteoarthritis. The course cannot be followed in a single patient but a possible sequence of events can be shown in a series (Figs. 1 to 6). The clinical evidence suggests that chondromalacia is less likely to progress when, at the time of diagnosis, it involves only a small area of the superficial and intermediate layers of the articular cartilage, but the greater the extent and severity of the changes, the greater the risk of osteoarthritis.

PATHOLOGY

Naked-eye appearances—Chondromalacia of the patella begins as a nodular swelling of part of the articular cartilage. It is situated more often on the medial than the lateral facet, commonly just below the centre. The nodular area is lustreless and grey or yellow in colour, and it is distinct from the glistening, blue, normal cartilage surrounding it. Fissures develop in the nodular zone and radiate from it, splitting the cartilage into irregular flakes (Fig. 2). The free edges of the flakes project above the surface of the surrounding articular cartilage. The fissures are usually directed obliquely into the depths of the cartilage so that the flakes, when viewed from the surface, do not stand upright but lie like the tiles of a roof, partly overlapping each other (Fig. 14). They are firmly attached to the underlying bone.

Hirsch (1944) has demonstrated by elaborate methods that, at the stage of nodular swelling, the elasticity of cartilage in the areas of chondromalacia is less than that of the surrounding, undamaged cartilage. Some elasticity remains even when the cartilage is fissured, as can

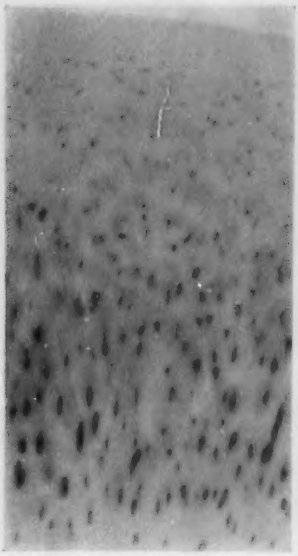

FIG. 7

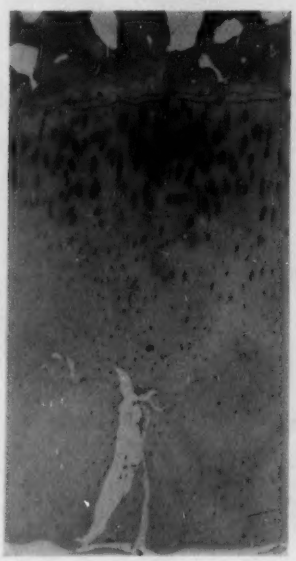

FIG. 8

FIG. 9

Figure 7—Normal articular cartilage of patella stained H. and E., ×25. Figure 8—Loss of normal staining in superficial and mid-zones, also fissure extending into mid-zone. H. and E., ×20. Figure 9—Irregular staining of superficial and mid-zone and cysts in the latter. H. and E., ×50.

FIG. 10

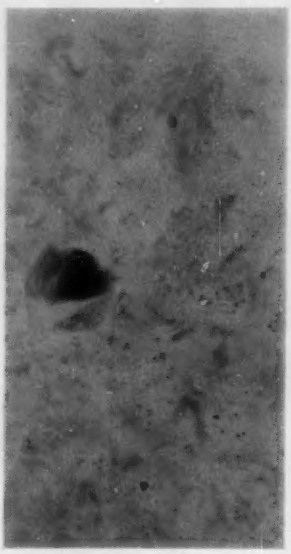

FIG. 11

FIG. 12

Figure 10—Hyperplasia of cartilage cells surrounded by areas of normal staining matrix. The line of calcified cartilage is broadened. H. and E., ×30. Figure 11—Loose body from knee joint of a case of chondromalacia showing cell nests and areas of normal staining matrix. H. and E., ×25. Figure 12—Late stage of chondromalacia showing degenerate cartilage with ossification and oedematous connective tissue in the marrow spaces. H. and E., ×30.

readily be shown by inserting a fine probe into a fissure and lifting the adjoining flake to an upright position; it snaps back into its original position when the probe is withdrawn.

The damaged cartilage, at first raised above the level of the surrounding articular surface, gradually becomes thinner and the flakes acquire a shaggy appearance (Fig. 3). They are then less elastic and softer, so much so that small pieces of frayed cartilage can be detached by rubbing with a finger. On opening knees with cartilage changes such as these, it is common to find cartilaginous loose bodies with rounded edges lying free in the joint.

When the condition is progressive the size of the damaged area increases and involves most of the medial facet, the ridge and the lateral facet, usually in this order. The cartilage becomes so thin that the underlying bone is exposed, giving an appearance rather like an " ulcer " surrounded by a rim of frayed cartilage (Fig. 3). The area of exposed bone increases and eventually becomes eburnated and often scored in a vertical direction (Figs. 4 to 6).

Osteophytes develop at the margin of the patellar cartilage where it joins the synovial membrane and capsular tissues. They appear soft, as rounded masses of bone covered by a thin, grey-white layer of cartilage. They do not occur in the nodular phase, but later when the fissuring is deep or extensive and the cartilage is thin. This is the stage of osteoarthritis; it is reached by only a few people under the age of forty-five but thereafter it becomes increasingly common. There is a close correlation between the sites of the cartilage damage and osteophyte formation. The medial facet is the most frequent site of cartilage damage, and the medial border is the usual site of osteophyte formation. When the lateral facet is involved, osteophytes tend to form along the lateral border. The more severe the lesion the greater is the probability that osteophytes will develop; of ten patellae removed surgically for severe chondromalacia, nine had marginal osteophytes.

The patellar facets on the femur may show changes in the articular cartilage which are sometimes referred to as " mirror lesions." The medial part is more often affected than the lateral, the naked-eye appearances resembling those in the patella. Occasionally the patellar facets of the femur are involved although the patella is normal.

The synovial membrane shows some hyperaemia and proliferation in almost every case of chondromalacia. In a few, usually near an area of fissuring, a synovial pannus spreads over the outer rim of the articular cartilage (Figs. 1 and 2). This pannus consists of relatively vascular connective tissue covered by flattened synovial cells, and it is so firmly adherent to the cartilage as to be inseparable.

Histology—Normal articular cartilage is hyaline cartilage possessing three poorly defined layers (Fig. 7). The deepest lies on the line of calcified cartilage over the chondro-osseous plate, and it contains large cells arranged in vertical columns. The cells in the transitional layer run obliquely, and in the superficial layer they are parallel to the surface. The size of the cells decreases from the deeper zones towards the surface. Mitotic figures are not seen in any layer of normal cartilage. The matrix in the deep zone is plentiful and stains a deep blue with haematoxylin and eosin, but nearer the surface it stains less deeply and appears metachromatic (Fig. 7). No fibres are visible in normal cartilage examined by routine histological methods.

Nodular swelling, which is the earliest sign of chondromalacia, has a typical microscopical picture. There is no visible alteration in the cartilage cells, but the matrix is changed and when it is stained with haematoxylin and eosin the superficial and transitional zones appear pinker (Fig. 8). We have found, using special techniques such as the " periodic acid Schiff reaction " (P.A.S.), that there is a decrease of P.A.S. positive matrix in swollen areas; this indicates a decrease in the chondroitin sulphuric acid content. Hirsch (1944) and Matthews (1953), using biochemical methods, have made a similar deduction. It seems, therefore, that the earliest change in chondromalacia is an alteration in the chemical structure of the matrix.

Fissures—The earliest fissures run tangentially to the surface, but the deeper the fissures the

more vertical is their course (Fig. 8). The ground substance, when examined with dark ground illumination, shows a fine, web-like system of thin fibrils which encircle cartilage cells or groups of cells. The fibrils appear to be intact even when there is marked fissuring.

Benninghoff (1925) demonstrated by maceration of normal articular cartilage an orderly system of fibres arranged in arcades. Although the course of the fissures corresponds with that of " Benninghoff's fibres," we have not been able to demonstrate any such fibres in normal or degenerate cartilage of the patella either by silver impregnation or by examination with polarised light.

Thirty-one specimens of patellar cartilage removed at operation on account of chondromalacia have been examined and the depths of the fissures compared with the loss of metachromasia. The results are shown in Table III.

TABLE III

CORRELATION BETWEEN LOSS OF METACHROMASIA AND FISSURE FORMATION

Metachromasia	Fissure formation			
	None	Superficial	Half thickness	Full thickness
Slight loss	1	6	0	0
Moderate loss	0	5	12	3
Complete loss	0	0	1	3

Of four specimens with complete loss of metachromasia, deep fissures were present in three. Of the twenty specimens showing moderate loss of staining of the matrix, three had fissures reaching through the full thickness. There is, therefore, correlation between the altered staining reaction of the cartilage and the depth of the fissures.

Cysts of cartilage—Cysts are often present in degenerate cartilage when there is fissuring. They appear as irregular spaces, usually situated at the base of fissures, but sometimes nearer the surface (Fig. 9). Although a majority are empty, a few contain a homogeneous gelatinous substance which stains sometimes with haematoxylin and sometimes with eosin, but never with both. It does not, whether it is basophilic or eosinophilic, give a constant P.A.S. reaction. Cysts were present in twenty out of thirty-one surgical specimens; metachromic staining of the cartilage around them was reduced, slightly in six, moderately in thirteen and completely in one.

Hyperplasia of cartilage—In well established chondromalacia there are focal areas in which the cartilage cells show hyperplasia both by forming cell nests and by a return of the staining properties of the ground substance in and around the nests. The ground substance appears to be streaming around the cell nests (Fig. 10). The process can be interpreted as an attempt at regeneration, but it remains circumscribed and the continuity of the cartilage is not restored. The cartilaginous loose bodies found in chondromalacia behave in a similar way (Fig. 11).

Two specimens have been examined in which the articular cartilage had first been removed at operation and then, about a year later, the patella was excised. The articular surfaces of the patella showed only a few areas of regeneration, the new cartilage being fibrous rather than hyaline in structure. A similar appearance is often seen after " mould " arthroplasty of the hip, as first recorded by Smith-Petersen (1948).

Bone and basal cartilage—The reaction of bone underlying areas of chondromalacia is proliferative. The changes begin when fissures first appear in the cartilage and then the line of calcified cartilage increases in thickness (Fig. 10). As the fissuring extends into the cartilage the increase continues until the thickness of the line is doubled or trebled. New bone is formed in the basal zone and small blood vessels from the marrow spaces penetrate the layer of

calcified cartilage. There is no evidence of ischaemic necrosis in the bone either before or after the cartilage is eroded.

A comparable reaction takes place at the edge of the patella. The surface cartilage proliferates, producing cartilage of a fibrous type, ossification occurs in the deeper layers, and marrow spaces form in the new bone. Thus are formed expanding nodules of fibrocartilage which ossify in the deeper parts to become osteophytes. This process occurs near areas of advanced cartilage degeneration and it also can be interpreted as an effort at regeneration. Occasionally proliferation occurs at the centre of a facet (Figs. 15 and 16).

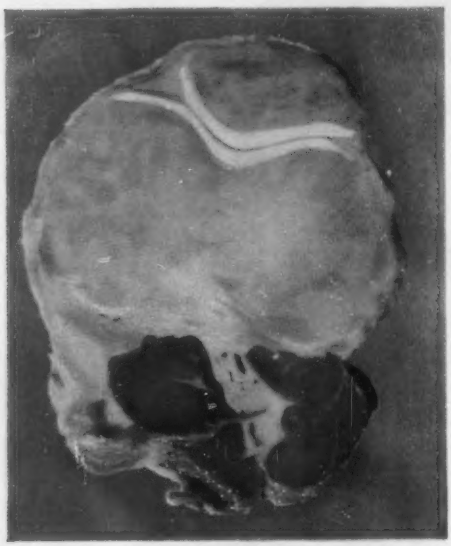

FIG. 13
Section of a knee flexed 75 degrees showing the extent of the congruity of the articulations between the medial and lateral patellar facets and the femur.

Bone cysts—The bone underlying areas in which the cartilage has been destroyed becomes sclerosed, and small cystic spaces filled with oedematous connective tissue appear beneath the surface (Figs. 12 and 16). These cysts have stomata communicating with the joint, but we have not seen any detritus from the joint inside them.

ETIOLOGY

The earliest demonstrable change in chondromalacia of the patella is an alteration in the reaction of the cartilage matrix to staining. This is due not to oedema, as many authors have maintained, but to a decrease in the chondroitin-sulphuric acid content. It begins in the lower part of the medial facet of the patella, which is said by Øwre (1936) to be the thickest articular cartilage of the body. The reason for this localisation has been investigated by Wiberg (1941). He cut slices of normal knee joints frozen at different angles of flexion and showed that, in the flexed knee, congruity between the medial articular surfaces of the patella and the femur was always less than between the lateral articular surfaces.

We have confirmed Wiberg's observation using a rapid-freeze technique (Fig. 13). The lateral facet of the patella is concave in two planes and makes contact with the femur through

FIG. 14

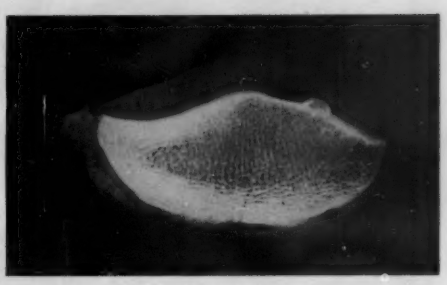

FIG. 15

Figure 14—Fissuring of the inner and lower quadrant of the articular cartilage. Figure 15—An osteophyte is developing in the middle of the lower and inner quadrant beneath fissured and partially eroded cartilage.

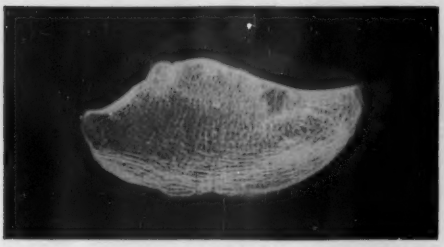

FIG. 16

FIG. 17

Figure 16—There is a cyst in the bone at the outer side, and an osteophyte is forming near the median ridge. Figure 17—The entire surface of the cartilage is rough and an osteophyte is forming at the outer margin.

much of the range of movement. The medial facet is, however, slightly convex and it touches the convex surface of the femur only at a single point, except when the knee is flexed less than 60 degrees. The lower part of the medial facet makes even less contact than the upper part, and it has been suggested by several writers that chondromalacia is caused by mechanical stress applied to this surface. The supporting evidence is not strong.

Harrison, Schajowicz and Trueta (1953), when studying osteoarthritis of the hip, found that there was more damage to the articular cartilage at the non-weight-bearing than at the weight-bearing areas of the femoral head. They suggested that normal movement with its alternating pressure and rest exerted a pumping action which helped to maintain the nutrition of articular cartilage. The patella is not a weight-bearing surface, and slab radiographs do not show a trabecular pattern delineating stressed and non-stressed areas of bone (Figs. 14 to 17). Nevertheless the localisation of the lesions in the patella supports the thesis that there is an association between pressure, or its absence, and the evolution of chondromalacia. Whether this is so or not, it is our view that trauma is of importance more often as an aggravating than as a primary factor, and that it acts by disrupting already degenerate cartilage. The final stage of osteoarthritis is regarded as the result of changes in cartilage, bone and synovial membrane taking place as a reaction to degeneration of the cartilage and to the liberation of free particles of degenerate cartilage within the joint.

CLINICAL FEATURES

Symptoms—The patient is commonly a young adult or in early middle age, but symptoms may occur during adolescence and occasionally in childhood. The onset is often insidious, but it may be precipitated by a minor injury such as a blow or a twist which is regarded by the patient as the cause. Occasionally there is a history of a more violent injury several months

before the symptoms begin. Some disability may have been present for months or years, and advice is sought only after a sudden exacerbation.

When the onset is insidious there is at first discomfort rather than pain; it is noticed after exercise and is not well localised. There may also be discomfort after sitting for some time, when first moving afterwards, or when going down stairs. Later the feeling of discomfort is replaced by pain, and there is often some swelling from effusion. The symptoms are, however, intermittent and, since the knee returns to normal after a couple of days, games such as football have not been given up. At a more advanced stage, which is reached by comparatively few, the pain and swelling become constant, with exacerbation after stress. There is considerable disability, and most games and the heavier types of work are impossible.

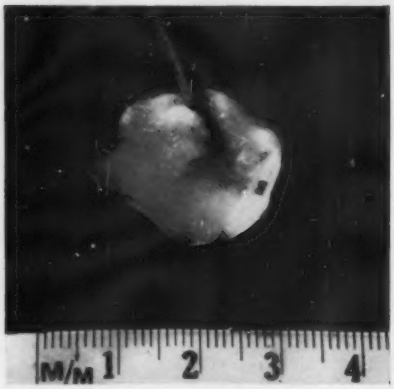

FIG. 18
Case 16—The articular cartilage after removal showing a large flap.

A history of "catching" of the knee during flexion or extension, but seldom both, is common. The patient may say that the knee lets him down. The "catch," which is produced by a partly detached flake of cartilage (Figs. 1, 3 and 18), is felt during flexion when the lower end of the flake is loose and during extension when the upper end is free. It may be more noticeable when a little pressure is applied to the patella during movement. True locking occurs when there is a loose body in the joint, as is quite common in chondromalacia.

The history can be summarised as one of intermittent discomfort or pain which is worse after sitting or after exercise, perhaps accompanied by a moderate effusion and sometimes by a "catch" during movement. There may be a spontaneous remission, or mild symptoms may continue indefinitely without getting worse. Sometimes there is an exacerbation associated with injury; sometimes there is gradual deterioration until finally the symptoms merge with those of osteoarthritis.

Physical signs—A positive diagnosis of chondromalacia can be made by the physical signs, but neither the history nor the signs give a reliable indication of the severity of the pathological changes. The area of cartilage involved can usually be determined, but not the stage to which degeneration has progressed. There may be considerable disability and marked tenderness beneath the patella when there is no more than a little fissuring of the cartilage, or the cartilage may be in a state of advanced disintegration when both symptoms and signs are relatively slight.

A variety of physical signs have been described but several are of little diagnostic value: patello-femoral crepitus on active movement of the knee, pain caused by rubbing the peripatellar tissues against the edge of the bone, effusion, and enlargement of the infrapatellar fat pads may be present but are open to other interpretations. Patello-femoral crepitus is particularly misleading. The important physical signs are:

1) *Pain* on grating the patella against the femur. With the knee extended, the muscles are relaxed and each quadrant in turn is gently rolled on the femur. The relation of the patella to the articular surface of the femur varies, and it may be necessary to push the patella downwards to ensure that all its articular surface comes into contact with the femur.

Pain may also be caused by pressure on the patella while the joint is moved actively or passively; as a rule it is felt only at certain points on the arc of movement.

2) *Tenderness* on pressure over the articular surface of the patella. This is accessible to palpation when the bone is displaced first to one side and then the other (Fig. 19). The amount

it can be displaced varies with the individual but often the whole under-surface except the median ridge can be felt, and even that is exposed when the capsule is very lax.

3) *A "catch"* on flexing or extending the knee can often be felt when there is a partly detached flake of cartilage (Figs. 1, 3 and 18). It may be easier to detect when the patella is gently pressed against the femur, and it always occurs at the same point on the arc of movement.

The presence of pain on grating the patella, together with tenderness beneath the patella, are diagnostic of chondromalacia provided other diseases such as rheumatoid arthritis have

FIG. 19
Examining for tenderness beneath the patella.

been excluded. Both signs must be present and at exactly the same part of the patella. Care must also be taken to ensure that the natural discomfort of the examination has not been misinterpreted by the patient. At operation the area of cartilage involved is often found to be greater than the physical signs suggested, but only occasionally less.

Pain and tenderness are usually severe when a vascular pannus covers part of the articular cartilage (Figs. 1 and 2). But when there is no pannus, an explanation of these signs is not easy to find because articular cartilage contains no sensory nerve endings, nor have we been able to demonstrate them in abnormal cartilage. An obvious suggestion, so far as tenderness beneath the patella is concerned, is that the synovial membrane interposed between the finger and the cartilage is inflamed. However, the same area of synovial membrane is seldom tender when pressed against the femur, and at subsequent operation it usually appears to be normal. Nor is the tenderness in the peripatellar synovial fold (although this, when inflamed, causes tenderness at the margin of the patella) because it cannot be invaginated far enough.

Radiographic examination—Radiographs are of assistance only in so far as they exclude other disease. In early and moderately advanced chondromalacia the bone is not involved and the thickness of the cartilage is not greatly reduced, so both the bone and the "joint space" appear normal. We have made many attempts to demonstrate the articular cartilage radiologically using every projection we could think of, both with and without the aid of air and other contrast media, but only on rare occasions have they been successful.

DIFFERENTIAL DIAGNOSIS

A positive diagnosis of chondromalacia is, in our experience, nearly always correct, but even when one is well aware of the condition and is constantly on the watch for it, it is surprising how often it is thought to be something else. The common mistake is to call

chondromalacia a tear of the medial semilunar cartilage, and next in frequency is the failure to recognise it as a cause of loose bodies. The association of chondromalacia with recurrent dislocation of the patella is too well known for the former to be overlooked (Macnab 1952); but it is not so well known that the patella is often affected when there is osteochondritis dissecans of the femur. Systemic diseases such as rheumatoid arthritis are unlikley to cause confusion provided reasonable care is taken.

Injuries of the medial semilunar cartilage—Most experienced surgeons will admit an appreciable error in the diagnosis of lesions of the semilunar cartilages. We have made this mistake nine times during the six years 1950 to 1955, an error of five per cent, and on each occasion there has been chondromalacia sufficiently advanced to account for the symptoms. The difficulty arises chiefly when there is a history of a twist, perhaps at football, which precipitated the onset of symptoms, and when there is complaint that " the knee lets me down," especially if there is also tenderness caused by detached fragments of cartilage that have fallen to the lowest part of the joint. In these circumstances, if the patellar pain and tenderness are but slight, it is tempting to incriminate the semilunar cartilage.

Loose bodies—Detached flakes of articular cartilage so often grow in size and calcify in the centre that chondromalacia should be suspected when there are loose bodies in the knee, whether or not there is radiological evidence of osteochondritis dissecans of the femur.

TREATMENT

The results of operation can be discovered by the ordinary methods of follow-up, but the difficulty in assessing conservative measures is that the precise condition of the articular cartilage is usually unknown. Neither the symptoms nor the signs are a reliable guide to the extent of degeneration. Moreover, in the absence of treatment, deterioration in some knees is rapid, in some it is slow, and others appear to recover. Until there is a clinical method of gauging accurately the condition of the cartilage and of following its course in individual patients the prognosis must remain uncertain and treatment can only be *secundum artem*.

The problem of treatment may be presented to the surgeon in several ways: 1) after a clinical diagnosis of chondromalacia, or of chondromalacia associated with another disorder; 2) on discovery of chondromalacia at an operation performed for another condition that has been misdiagnosed, the symptoms really being due to chondromalacia, or for another condition that proved to be present, the chondromalacia having caused no symptoms, or only trivial ones.

A clinical diagnosis may be made at any stage in the course of the disease because some patients present themselves with trivial symptoms, whereas the more robust wait until they are incapacitated. There is an infinite variation from trivial through moderate to severe. Treatment is hardly in doubt at the extremes. With minor symptoms and correspondingly slight physical signs it must be conservative. When the symptoms are incapacitating and long continued, operation gives the best chance of restoring function. The intermediate grades should first be treated conservatively and operation considered only if there is continued deterioration. There is insufficient information as to which knees will ultimately develop osteoarthritis to justify operation for this reason alone. It is our practice to advise operation only when it is indicated by the severity of the disability. Excision of the patella usually gives a good or a fair result, but seldom a perfect one, and there must be a reasonable prospect that this operation will leave the patient with a better knee than he already has.

Discovery at operation offers rather a different problem because the exposure has already been made and the condition of the cartilage can be seen.

Small areas of swollen or soft cartilage are probably best left untouched, but if there is fissuring the affected cartilage may be shaved off; it is usually unnecessary to remove the whole thickness. Lesions of this sort will be found quite often if the under-surface of the patella is inspected at every operation. Areas up to about one inch in diameter can be shaved

after slightly enlarging the incision for meniscectomy; the patella need not be everted and the time of recovery is not materially prolonged.

Advanced chondromalacia that has clearly been causing symptoms should be dealt with in the same way as if the operation were one of choice. In the absence of previous permission it may, however, be inadvisable to excise the patella.

Conservative treatment—There is no evidence that conservative measures have any effect on the course of the complaint. Many patients with mild symptoms improve without treatment, and many improve with it. However, the clinical impression is that treatment is responsible for some amelioration. During the last three years we have treated conservatively upwards of one hundred patients. Those with trivial symptoms have seldom attended for longer than a few weeks, but when the disability has been greater, they have often thought it worth while to continue for months. The effect on the subsequent course will not be known for some years.

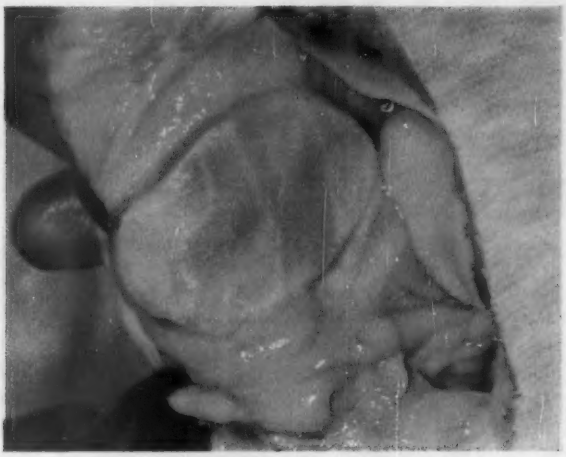

FIG. 20
Case 28 after removal of nearly all the articular cartilage of the patella.

The methods of treatment available are rest, short-wave diathermy, and intra-articular injections. Diathermy is prescribed in every instance. Rest, when the symptoms are trivial, may consist only in avoiding games and all exercise that is not essential, but occupations involving complete and powerful extension of the knee, such as ballet dancing and walking races, should be stopped. With more severe symptoms immobilisation is desirable and a bivalved plaster-of-Paris (or polythene) cylinder is applied with the knee flexed 15 degrees. It is taken off at night, and to facilitate diathermy. Exercises without resistance may be given to reduce the amount of wasting of the quadriceps, but exercises against resistance are contra-indicated. We have no experience of intra-articular injection of hydrocortisone for this complaint.

Operation—Two procedures have been practised—removal of the affected articular cartilage and excision of the patella. Patelloplasty has not been attempted because our experience with this operation performed for osteoarthritis has not been sufficiently encouraging.

The patella is first inspected through a medial parapatellar incision, and if extensive changes are seen the incision is prolonged to allow the bone to be everted. The complete incision begins at a point in the mid-line one and a half inches above the patella, and extends downwards for an inch, splitting the quadriceps. It then curves round the inner side of the bone, dividing the musculo-tendinous junction and approaches the mid-line again at the level of the tuberosity of the tibia.

Extensive fissuring, flaking and erosion of a considerable area of the articular cartilage is an indication to remove it, but whether this is best effected by shaving off the affected cartilage or by excising the patella is not yet established.

Removal of cartilage—This has been our practice when the area involved has not been greater than about one-third of the whole. With more extensive involvement we have removed the affected cartilage, if necessary, from the entire patella, when chondromalacia has been a chance finding and permission to excise the patella has not been obtained. It has also been done on other occasions after previous discussion with the patient.

TABLE IV

METHOD OF TREATMENT IN SIXTY-FIVE KNEES WITH CHONDROMALACIA OF THE PATELLA SEEN AT OPERATION

	Males	Females	Total
Seen at operation, not treated . .	14	5	19
Part or all articular cartilage removed	20	11	31
Patella excised	7	8	15
Total number of knees . . .	41	24	65*

* The number of patients concerned is sixty-two.

Nodular, soft cartilage should be removed as well as fissured. This is done by shaving with a scalpel, the healthy cartilage being bevelled at the margin to leave a smooth surface. The bone is not usually exposed but remains covered with the basal layer of cartilage (Fig. 20). On a few occasions a small area of the femur has been treated in the same way, but extensive affection of the femoral part of the joint is an indication for excision of the patella. Post-operative recovery is often rather slow, and although function may be restored sufficiently to permit light work after a month, physiotherapy may be necessary for much longer.

Excision of the patella—This operation gives short-term results similar to those obtained when it is performed for fracture. Function is seldom restored completely to normal, but the risk that a second operation may later become necessary is removed. The appearance of the knee is usually quite good, but occasionally the contour of the knuckle of the femur becomes obvious and this is most unsightly.

The patella, in younger people, can usually be dissected out without injuring the quadriceps expansion to such an extent that it needs repair. In older patients more fibres are inserted into the central part of the bone and it may become so attenuated that suture is desirable. Restoration of function often takes rather longer than after removal of the articular cartilage, and manipulation may be necessary to restore full flexion.

RESULTS

We have records of the condition of the articular cartilage as seen at operation in sixty-five knees with marked chondromalacia. Those with frank osteoarthritis are deliberately excluded from this survey.

Reference has already been made under the heading of natural history to the nineteen knees in which the cartilage was left untouched. The remaining forty-six were treated by removal of all or part of the articular cartilage, or by excision of the patella. The particulars are detailed in Table V and the results are summarised in Table VI.

The result has been called good when the patient is able to follow his usual occupation and lead an ordinary life without discomfort. The knees may not be perfect but they do stand up to any ordinary strain. For example, the patient in Cases 1 and 26, who had articular

TABLE V

OPERATIVE TREATMENT OF CHONDROMALACIA (TO APRIL 1955)

Case number	Age	Sex	Occupation	Pre-operative diagnosis	Date of operation	Time to "normal" (*weeks*)	Date last seen	Result	Comment
ONE-THIRD OF ARTICULAR CARTILAGE REMOVED									
1	22	M	Sanitary engineer	Chondromalacia	Feb. 1952	16	May 1955	Good	Operation on both knees. This one causes no trouble. See Case 26
2	16	M	Bank clerk	Osteochondritis dissecans: Chondromalacia	Oct. 1953	22	Aug. 1955	Good	Loose fragment removed from femoral condyle. Now has no disability
3	16	F	Clerk	Chondromalacia	Dec. 1953	8	Oct. 1954	Good	Obese. Now pupil nurse and knee causes no trouble
4	37	M	Sales manager	Chondromalacia	May 1954	13	June 1955	Good	Superficial scar tender. No other symptoms
5	41	M	Heavy lorry driver	Chondromalacia. Torn medial semilunar cartilage	June 1954	—	—	Bad	Torn semilunar cartilage removed. Patella excised four months later for continuing pain and disability. (See Case 44)
6	21	M	Metal machinist	Chondromalacia	Aug. 1954	14	Aug. 1955	Good	Now has no symptoms
TWO-THIRDS OF ARTICULAR CARTILAGE REMOVED									
7	15	M	School	Chondromalacia	Mar. 1952	—	—	Bad	Patella excised eight months later. Other knee also affected. See Case 35
8	19	F	—	Torn medial semilunar cartilage	Sept. 1953	8	Oct. 1954	Good	Semilunar cartilage normal. Now no symptoms
9	16	F	School	Torn medial semilunar cartilage	Oct. 1953	50	Aug. 1955	Good	Slow to recover but now free of symptoms
10	27	M	Doctor	Multiple loose bodies	Nov. 1953	10	Oct. 1954	Good	More than 100 cartilaginous loose bodies removed. Now has occasional ache
11	24	M	Industrial chemist	Torn medial semilunar cartilage	Nov. 1953	28	May 1954	—	Semilunar cartilage normal. Now in South Africa
12	21	F	P.T. instructor	Torn medial semilunar cartilage	Apr. 1954	8	Sept. 1955	Good	Now practically normal
13	45	M	Flat manager	Chondromalacia	May 1954	50	June 1955	Good	Slow to regain good function but now has only slight swelling after much exercise
14	23	M	R.A.F.	Torn semilunar cartilage	June 1954	8	May 1955	Good	Semilunar cartilage normal. Now has occasional " stiffness "
15	18	M	Student	Torn medial semilunar cartilage	June 1954	6	Aug. 1955	Good	Torn semilunar cartilage removed. Now free of symptoms but other knee beginning to cause trouble

TABLE V—*continued*

Case number	Age	Sex	Occupation	Pre-operative diagnosis	Date of operation	Time to "normal" (*weeks*)	Date last seen	Result	Comment
TWO-THIRDS OF ARTICULAR CARTILAGE REMOVED—*continued*									
16	28	M	School and games master	Torn medial semilunar cartilage	Aug. 1954	12	June 1955	Good	Sometimes swells after much exercise
17	43	M	Inspector of taxes	Torn cruciate and medial ligament	Aug. 1954	—	June 1955	Bad	Repair of medial ligament; meniscectomy. Knee deteriorating and swells after moderate activity
18	20	M	Student	Torn medial semilunar cartilage	Jan. 1955	8	Mar. 1955	—	Free of all symptoms two months after operation
19	37	M	Garage hand	Loose bodies	Mar. 1955	—	June 1955	—	At work but still lacks 25 degrees flexion
20	28	M	Clerk	Loose body and osteochondritis dissecans	May 1955	—	—	—	Articular cartilage removed from femur also
21	22	M	Librarian	Chondromalacia	May 1954	6	May 1955	Good	Knee sometimes feels stiff
22	33	M	Telephone engineer	Torn medial semilunar cartilage	June 1955	—	—	—	Torn semilunar cartilage
ALL ARTICULAR CARTILAGE REMOVED									
23	—	F	Housewife	Torn medial semilunar cartilage	Dec. 1951	16	Aug. 1954	Good	Only symptom is aching in wet weather
24	27	F	Secretary	Chondromalacia	Aug. 1952	—	Feb. 1954	Fair	Gross villous synovitis. Very slow recovery but now is usually free of symptoms and can walk miles; occasional episodes of stiffness and swelling
25	15	F	Telephonist	Torn medial semilunar cartilage	June 1953	—	—	Bad	Subsequently had patella excised elsewhere
26	25	M	Sanitary engineer	Chondromalacia	Mar. 1954	12	June 1955	Good	Operation on both knees. No trouble except some pain after running about twelve miles. See Case 1
27	22	F	Nurse	Torn medial semilunar cartilage	May 1954	12	July 1955	Good	Semilunar cartilage normal. Only symptom is some aching after kneeling for long
28	36	F	Technical staff B.B.C.	Chondromalacia	Sept. 1954	27	Aug. 1955	Good	Now has no symptoms
29	43	F	Canteen manageress	Chondromalacia	Oct. 1954	—	July 1955	Bad	Still much disability; requires excision of patella
30	28	M	Transport executive	Chondromalacia	Feb. 1955	6	July 1955	Good	Only symptom is swelling after several sets of tennis
31	24	F	Secretary	Chondromalacia	Apr. 1955	—	July 1955	—	No pain but has not yet regained full flexion

TABLE V—*continued*

Case number	Age	Sex	Occupation	Pre-operative diagnosis	Date of operation	Time to "normal" (*weeks*)	Date last seen	Result	Comment
PATELLA EXCISED									
32	34	F	Clerk	Chondromalacia	June 1948	16	May 1955	Good	Aches if " overdoes it "
33	32	M	Air steward	Chondromalacia	Feb. 1950	—	June 1950	—	Cannot be traced
34	25	M	Salesman	Chondromalacia	Aug. 1950	?	Sept. 1954	Good	Now has no symptoms
35	15	M	School	Chondromalacia right patella.	Nov. 1952	20	Sept. 1954	Fair	Previous removal two-thirds cartilage right patella (Case 7). Now signwriter. Both knees ache if he stands long, and does not like climbing ladders
36	16	M	School	Chondromalacia left patella	Mar. 1953	?	Sept. 1954	Fair	Previous removal two-thirds cartilage right patella (Case 7). Now signwriter. Both knees ache if he stands long, and does not like climbing ladders
37	17	F	Shop assistant	Recurrent dislocation with chondromalacia	Jan. 1953	60	May 1955	Fair	Transplantation patella tendon. Slow regaining function. Now finds kneeling difficult and sometimes swells
38	34	F	Housewife	Chondromalacia	Sept. 1953	24	Sept. 1954	Good	Aches after sitting for long
39	24	F	Housewife	Chondromalacia	May 1954	No	July 1955	Bad	Several previous operations on same knee elsewhere. Unlikely ever to get good function
40	33	F	Saleswoman	Chondromalacia	June 1954	—	Aug. 1955	Good	Lacks 10 degrees flexion. Aches in cold weather
41	27	M	Engineer	Chondromalacia	Aug. 1954	24	June 1955	Fair	Lacks 50 degrees flexion and has difficulty in kneeling
42	28	F	Invoice clerk	Chondromalacia; old recurrent dislocation	Aug. 1954	12	Apr. 1955	Good	Transplantation patellar tendon 1945. Now has no symptoms
43	40	F	Hotel manageress	Chondromalacia; loose bodies	Oct. 1954	—	May 1955	Fair	Still swells after exercise
44	42	M	Heavy lorry driver	Chondromalacia	Oct. 1954	12	Aug. 1955	Fair	Still aches and occasionally gives way. Driving a light van. See Case 5
45	19	F	Teaching student	Chondromalacia	Apr. 1955	—	July 1955	—	Much muscle wasting before operation and has not yet gained full control
46	49	M	Clerk	Chondromalacia	Apr. 1955	—	July 1955	—	Back at work but has not yet full flexion

cartilage removed from both patellae, has had to give up his hobby of long-distance running because the knee that had all the cartilage removed hurts after about twelve miles.

Removal of cartilage—Thirty-one knees have been treated in this way, nineteen being classified as good, one as fair, five as bad, and six are recent or unknown. Only one has been called fair because a moderate early result usually deteriorates quickly and becomes bad. The exception is the patient in Case 24 who had a gross villous synovitis and has been very slow to recover. Three years after the operation she is unconscious of her knee most of the time, and usually

can walk miles without any trouble, but occasionally there are episodes of stiffness and swelling lasting a few days. The patients with bad results have either had the patella excised later or are likely to require further operation.

The results become less satisfactory as the amount of cartilage removed increases. However, the bad results include two patients aged forty-three, and two with very extensive degeneration at the age of fifteen. The older patients, who are approaching the age of osteoarthritis, are probably better treated by excision of the patella. Very young patients with extensive degeneration are likely to have early involvement of the femoral articular cartilage and a lasting result is improbable whatever is done. Better selection of cases for removal of cartilage should improve the results.

TABLE VI

RESULTS OF OPERATION (FORTY-SIX CASES)

Nature of operation	Result				
	Good	Fair	Bad	Recent or unknown	Total
Articular cartilage removed:					
one-third	5	—	1	—	6
two-thirds	9	—	2	5	16
completely	5	1	2	1	9
Patella excised	5	6	1	3	15
Total	24	7	6	9	46

The only comparable figures are those given by Karlson (1940) who re-examined the patients operated upon in Aleman's clinic one to twenty years before, but he did not give details of the amount of cartilage removed. Of forty-eight knees with chondromalacia (but without frank osteoarthritis), thirty-two had made a functional recovery, thirteen had improved and three had not improved; seven patients (it is not stated in which group) had radiological evidence of osteoarthritis.

Excision of the patella—The patella has been removed for chondromalacia on fifteen occasions. The results are not strikingly good but they compare with those of the corresponding operation for recent fracture, which are not as brilliant as is commonly supposed. Scott (1949) followed up a number of men who had been operated upon in the Royal Air Force during the war. He found that only 5 per cent regarded themselves as having normal knees; the remainder had a variety of troubles, for example, 79 per cent complained of pain and 55 per cent said that they were unable to walk downstairs normally.

The long-term results of the more conservative operation will not be known for some years. If complete removal of all degenerate cartilage prevents the reactive changes that lead to osteoarthritis, it may well become the procedure of choice in properly selected cases.

SUMMARY

1. Chondromalacia, sometimes a precursor of osteoarthritis, is present in the articular cartilage of the patella of most people by the age of thirty; it causes symptoms in only a few, and it gives rise to osteoarthritis in fewer still. It may progress slowly or quickly but there is no clinical method of assessing the prognosis at an early stage.

2. The earliest change is swelling of the cartilage associated with a decrease in the chondroitin sulphuric acid content of the matrix. Later the cartilage fissures and flakes off to expose the bone, and there are reactive changes in the cartilage, bone and synovial membrane. The process is described and the etiology discussed.
3. The symptoms, signs and treatment are discussed. Operation, which has been performed only when there are disabling symptoms, may consist in removing part or the whole of the articular cartilage, or in excision of the patella. The results in forty-six knees are given.

It is a pleasure to thank Mr M. Turney for his help with the many photographs, and Mr P. Runnicles for the photomicrographs.

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SUBOCCIPITAL ARTERIOVENOUS ANEURYSMS OF THE VERTEBRAL ARTERY

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The fact that arteriovenous fistulae of the suboccipital portion of the vertebral artery are very uncommon is made clear by the real scarcity of cases in the literature. The reasons for this can be conjectured to be twofold: 1) the chances are small that a missile will penetrate deeply enough to injure the vertebral artery without at the same time inflicting irrecoverable damage on the highest segments of the cord or even the medulla oblongata; and 2) even though the conditions requisite for survival be fulfilled, the chances are still inconsiderable that precisely the correct lesion will be inflicted on the vertebral artery to cause a fistulous communication into the regional veins.

Two examples will be presented of arteriovenous aneurysms of the vertebral artery in its horizontal or atlantal portion cured by operation—both our own cases. A third case, of arteriovenous aneurysm between C.1-2, has been kindly added by Professor Norman Dott. Opportunity will be taken to describe the operative steps necessary for the cure of such aneurysms, a subject of which there is something still to be said in spite of the careful studies made by several German surgeons after the first world war, such as Küttner (1917) and Drüner (1917). Since the presently reported cases were undertaken and certain lessons learned, the great series treated by Elkin and Harris (1946) has been published with reflections on the operative handling of this difficult kind of aneurysm. Before this Henry (1927) had described an approach to the vertebral artery in that part which lies between the transverse processes of atlas and axis, the end of the so-called second part. Henry's operation is not strikingly different from Drüner's. The subject is one which can be carried further, not only with regard to proximal ligature, which is insufficient for a fistulous aneurysm though it seems to have succeeded for saccular, but as regards the relations of the vessel in a region where it is very rarely exposed in the living subject. A further matter of importance is the distribution of the veins in relation to the vertebral artery. This vessel is usually depicted as bare, with no accompanying venae comites. How therefore can it come about that an arteriovenous fistula is possible? The reason is, of course, that there are numerous veins (as has been uniquely well shown in Figure 6 which is reproduced from Grant's *Atlas* (1951) by courtesy of the author) connecting with the important profunda vein which runs down the neck deep in the muscles. Since these veins have not been thought to be of much clinical significance they have commonly been ignored in illustrations. A study of these matters has been made by one of us (R. A. B.), the results of which, together with a further presentation of the anatomy of approach, will be published later.

CASE REPORTS

Case 1—A soldier aged twenty was admitted to hospital in April 1945, where Mr Kerr diagnosed a suboccipital arteriovenous aneurysm. He was transferred for operation in May to the Neurosurgical Service of the Manchester Royal Infirmary. He had been hit by a bullet that traversed his head from a point on the left cheek medial to the infra-orbital foramen to its exit an inch from the nuchal furrow and two and a half inches below the inion. He had been given a short course of penicillin after a blood transfusion for shock. It was recorded that he had a haematoma in the palate. These wounds gave no trouble. The patient had

experienced immediate hoarseness when he spoke or coughed and had bled from the nose and mouth. He had difficulty in swallowing and in chewing and was still occasionally having attacks of dyspnoea and was still hoarse on admission. These disabilities had been due in the early days to local swelling but later to persisting cranial nerve damage which will be further detailed presently. There was weakness of the left shoulder. A short time after the injury the patient became aware of a noise in the head which he referred to the skull base and left ear, synchronous with the pulse. It became more definite and more obtrusive as time passed.

On admission his general condition was excellent. He was a tall, spare young man, fully ambulant and with small scars at the sites already indicated. The noise that he now believed to originate in his neck persisted. He was much less hoarse than he had been, and could swallow without difficulty. His complaints were of: 1) the bruit in his neck and head; 2) numbness of the left side of his tongue; 3) drooping of the left shoulder as he walked; 4) dyspnoea on exertion; and 5) inconsiderable pain in left face. High in the left side of the neck behind there was a diffuse and slightly red swelling elevated not more than one centimetre. On palpation a deep-seated thrill could be felt. On auscultation it had the characteristic sound of an arteriovenous aneurysm, the maximal intensity being half way between mastoid and inion and roughly level with the mastoid tip. This sound was somewhat diminished by carotid compression and also by turning the head strongly to the other side. This was verified with the stethoscope. The patient had himself observed that if he turned his chin far to the opposite side he could generally reduce the intensity of the bruit. A diagnosis was made of fistulous aneurysm either of the vertebral artery at some point in the suboccipital part of its course or possibly of the occipital artery.

Cranial nerve palsies—The patient presented signs of injury of several cranial nerves: 1) of the left lingual nerve; 2) there was relative numbness of the left faucies and the back of the tongue corresponding (glosso-pharyngeal nerve); 3) of the left hypoglossal nerve—the tongue was atrophied as well as numb; 4) of the left vagus, deduced from the history of hoarseness, difficulty in swallowing and dyspnoeic attacks; Professor Victor Lambert reported the left vocal cord paralysed (five months after the injury the movement of the left vocal cord was nearly normal again laryngoscopically); 5) weakness and some atrophy of both the left sternomastoid and trapezius pointed to a high injury to the spinal accessory nerve; 6) Horner's syndrome was present on the left, evidence of damage to the cervical sympathetic trunk or very probably of its superior ganglion. The patient therefore presented an extremely interesting group of cranial nerve injuries, not only those of the jugular foramen (known to the French as the Vernet " syndrome du trou dechiré posterieur," but with cervical sympathetic and hypoglossal added (the syndrome of Villaret). As has been mentioned, the hypoglossal and lingual branch of the trigeminal had also suffered damage. These cranial nerve injuries mapped out the course of the bullet and were interesting in themselves, but as they were not important to our present theme the subject will not further be pursued. The fundi were normal. Hearing was: wrist watch two inches left ear, fifteen inches right ear. Cold caloric test gave slightly delayed reactions on the left. The cerebro-spinal fluid pressure was normal and its constituents were normal. General condition: nothing of note; heart not enlarged; blood pressure 105/75. Circulation time was 55 seconds.

Operation—It was not known for certain whether the aneurysm was vertebral or occipital, though the former was the more probable because carotid compression had not eliminated it, and for the reasons given later in the general discussion. The significance of the reduction of the bruit by head turning probably, it was thought, favoured vertebral aneurysm.

In order to bring the whole region into view the operator (G. J.) decided to expose the carotid sheath, to divide the sternomastoid completely just below the mastoid process, and to carry the incision across the suboccipital region in such a manner that the occiput and atlas could be fully exposed. The patient was laid recumbent with a large sandbag under the left shoulder and the face turned well to the right. The skin incision was carried up the anterior

border of the sternomastoid and across it, dividing it at the mastoid tip to finish in the mid-nuchal line below the inion (Fig. 3). The carotid sheath, the vagus and spinal accessory nerves were quickly found, an advantage of very high section of the sternomastoid being that the accessory is in little danger. The vagus, accessory and hypoglossal were seen and the glossopharyngeal to the medial side high up where it wound round the stylopharyngeus. There

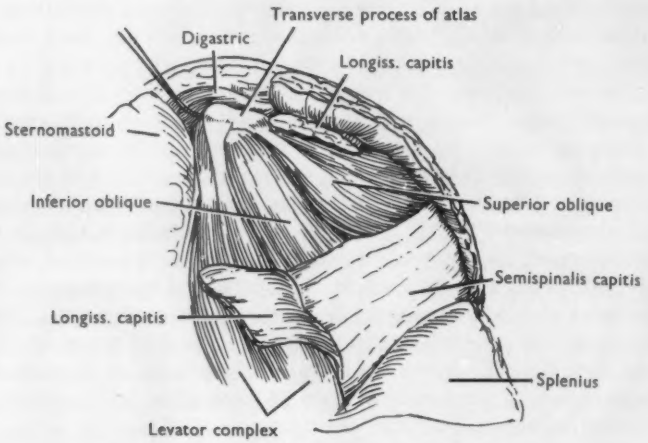

FIG. 1
Trapezius, splenius and longissimus capitis divided showing deeper muscles covering atlas vertebra.

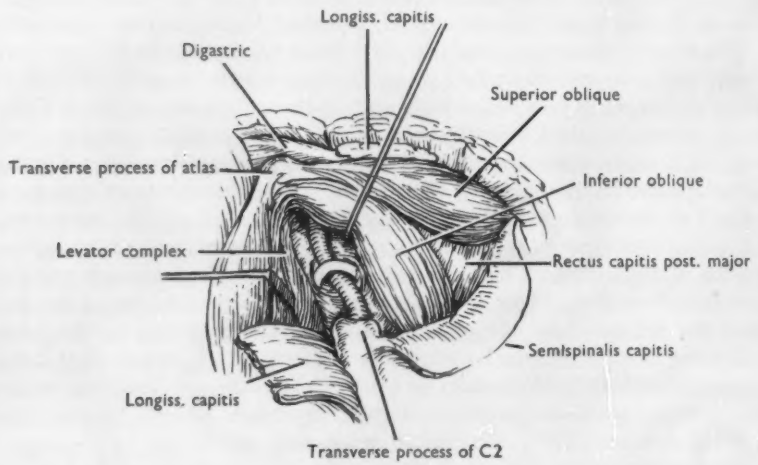

FIG. 2
Exposure of artery, crossed by second cervical nerve, between transverse processes of atlas and axis.

was some scar tissue surrounding them and the internal carotid artery, but the bullet track must have been very close to the skull base for the actual site of the nerve injuries was not visible. Since the nerves were, in the main, recovering there seemed to be no point in pressing the search. No trace of a carotid arteriovenous aneurysm was found. The incision was now deepened and the suboccipital muscles stripped from the occiput. There is no doubt that

the great familiarity with these steps that neurosurgery engenders was helpful at this point. The muscle clearance was carried down to the atlas as far as the mid-line by dividing the upper ends of trapezius, splenius and longissimus capitis, and later semispinalis and levator anguli scapulae. By the use of a boiled stethoscope used from time to time it was clear from the increasing loudness of the noise that the site of the fistula was being approached. The occipital squame was now almost bare but it required for its full lateral exposure the division of the belly of the superior oblique muscle running from the transverse process of the atlas to the occiput (Fig. 3). Dissecting in the gap between the superior and inferior obliques a large but thin-walled vessel was torn and spurted bright blood, evidently an efferent vein from the fistula. The transverse processes of the upper two cervical vertebrae had long been in view though their exact configuration was obscured by the muscles to them. However

FIG. 3
Photograph to show the incision used (Case 2).

the vertebral artery was eventually exposed below the transverse process of the atlas by division of the inferior oblique muscle. The vessel presented an unusual picture because the two loops instead of being at right angles to one another were almost parallel. It was later proved by R. A. Bailey's dissections that this unexpected appearance was due to the posture of the head, and that the lie of the artery was altered considerably by movement (Fig. 5). This change is important in the surgery of the upper reaches of the vertebral artery, but it is little to be wondered at that it has not excited comment because exposures in this situation have so rarely been made. At the time the appearance was disconcerting, but a review of the field of operation and a verification of visible landmarks left no doubt that this must be the artery we were seeking. The artery, where it rides away from the axis to enter the foramen in the atlantal transverse process, is very accessible and it was easily ligated with two double fine black silk threads. The bruit ceased, as was confirmed by palpation and the use of the sterilised stethoscope. Proximal ligature is notoriously an uncertain way of curing arteriovenous aneurysms, and there was a strong probability of the bruit's recurrence

in a few days. This should be a particularly likely event with the vertebral because the anastomosis with its fellow of the opposite side is at this level so very near. Accordingly the artery was dissected clear immediately before it pierced the posterior occipito-atlantal ligament above the arch of the atlas and two silver clips were used to occlude it. The fistulous communication was not further identified. The fistula was evidently in the horizontal part of the vessel's course between the two now occluded points. A careful search with the sterilised stethoscope proved that no trace of the murmur could be heard on direct contact with the deepest tissues. The wound was firmly closed in layers and healed without giving trouble. The patient was discharged a month later. There was no return of the bruit. He remains well ten years later.

Case 2—A man aged twenty-two was admitted to the Neurosurgical Service of the Manchester Royal Infirmary in October 1945 from a Royal Air Force hospital at the surgeon's request

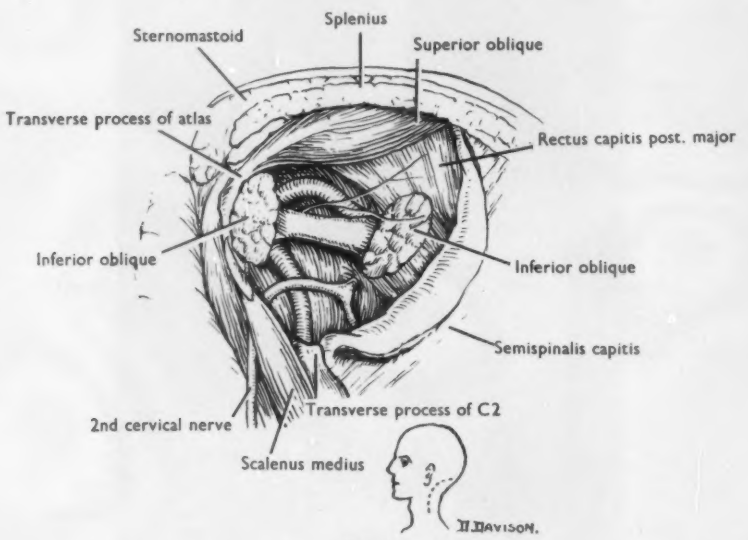

FIG. 4

Inferior oblique sectioned to show third part of left vertebral artery. In practice the superior oblique also requires division.

after consultation with Sir Stanford Cade. An unsuccessful attempt had been made to expose a left vertebral arteriovenous aneurysm in the suboccipital triangle.

The patient, a West Indian, had been stabbed deeply in the left suboccipital triangle with a sharp-pointed stick (the long slender shaft of a native " top ") six years before in the course of a schoolboys' squabble. He was taken to hospital but was not detained as there had been no noteworthy haemorrhage. The puncture healed without incident. About three weeks afterwards the patient became aware of what he described as a faint " gushing " noise heard in the left ear. It had continued during the ensuing six years but he had learned to ignore it. However it became louder as time passed and for the last eight months it had interfered with his sleep, though he was not otherwise inconvenienced. He had discovered for himself that deep pressure at a point behind his mastoid process stopped the noise. In September 1945 he had reported sick because of this noise and, a diagnosis of arteriovenous aneurysm having been made, two operations were done. A muscle flap was turned down in the left suboccipital region, but the aneurysm was not found. A few days later an external carotid angiogram failed to assist in the diagnosis. After this he was transferred to Manchester. There was a

horse-shoe scar commencing at the anterior border of sternomastoid and sweeping backwards into the suboccipital triangle. In the middle of this triangle some four centimetres below the superior nuchal line was the insignificant scar of the original punctured wound. The characteristic thrill and continuous hum of an arteriovenous fistula could be heard behind the mastoid tip and medial to it over the upper nuchal muscles. There was no notable swelling but deep pulsation could be felt. It was not forcible. The noise was not materially changed by carotid compression, and since the previous operation and angiography had not supported the idea of the aneurysm being connected with the occipital artery it followed that the aneurysm must be vertebral. It was again high and proved once more to be in the final or third transverse portion. In this case there were no cranial nerve palsies and no sign of any other nervous involvement. Electro-encephalography showed slight cortical suppression in the left parietal area but no neurological signs could be found to correspond. The patient

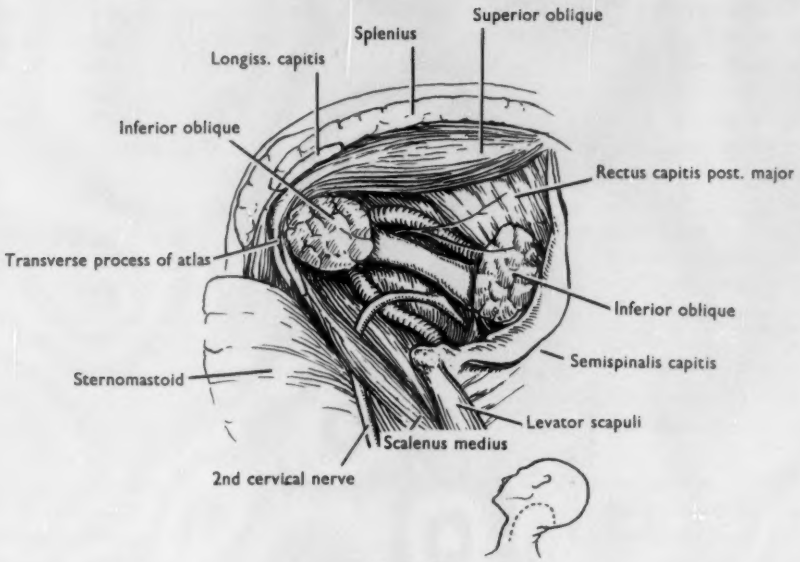

FIG. 5
Same as Figure 4 but head strongly turned to right altering direction of artery above and below transverse process of atlas.

complained of occasional vertigo and of slight headaches but he was a very nervous subject, who habitually construed all happenings as serious and threatening to his security. His electrocardiogram, after an arteriovenous fistula had been present for six years, showed only slight changes—slight depression of the RS-T segments in Lead III, probably within normal limits. The heart was not enlarged.

Operation—The old flap was reopened (G. J.), the sternomastoid divided at the mastoid tip and the muscles of the posterior triangle cut through down to the occipital bone. An excellent view of the carotid sheath was obtained and the absence of aneurysmal communication verified. The operative steps were not easy because of scarring from the previous exploration and shortly for another reason, for no sooner had the superior oblique been brought into view than a number of veins gave trouble. These veins were thin-walled, numerous and irregular (Fig. 6) and although no single vein was very large, there was an infiltration with scar tissue that made clean dissection impossible. Several of these veins were secured but in clearing the posterior arch of the atlas one or more were damaged and furious haemorrhage

resulted. It was stopped by finger pressure. As is usual with bleeding of this kind, it was difficult to define its source, which was somewhere on the arch of the atlas. It was later proved that the bleeding came from the vertebral artery or from an aneurysmal vein fused with it. It proved necessary for the operator to keep up his finger pressure for the next hour and a half, while the vertebral artery was exposed at what we may call the site of election between the transverse processes of atlas and axis by cutting through the levator mass and inferior oblique close to the atlas. This difficulty was incurred by failure to carry out a precautionary exposure of the length of artery between atlas and axis earlier in the operation. This may be a reasonably easy operation when an arteriovenous aneurysm does not impede dissection, but when there is already one source of haemorrhage that is only precariously controlled and a probability that a second accident will end fatally, it is an uncomfortable procedure. Eventually the

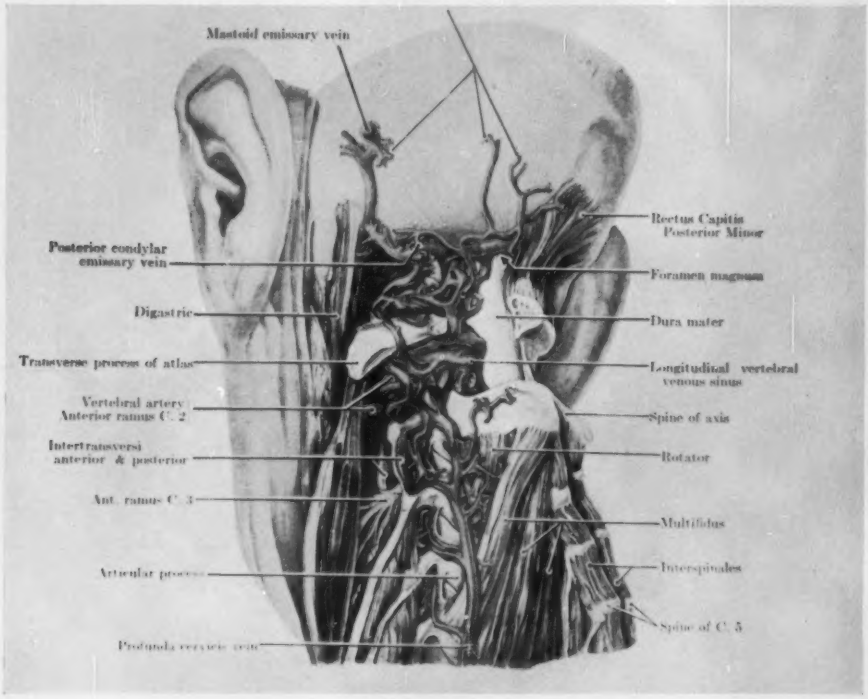

FIG. 6

The deep veins posteriorly of the neck. (By courtesy of Professor Boileau Grant.)

artery which presented as a loop was secured after some anxious dissection and after the transverse process of the axis had been nibbled away to get a better length of artery to work on. The second cervical nerve appeared to be stretched over the vessel, which was large, but there had been no symptom referable to such an arrangement. Once the vertebral had been ligated it became possible to relax the finger pressure sufficiently to allow the insertion of a number of black silk sutures to oversew the site of the aneurysm above the arch of the atlas. It was then seen that the source of the haemorrhage had been the vertebral artery itself at the point where it penetrates the occipito-atlantal ligament. Its entrance was sewn up. No trace of a normal artery could be found above the arch except laterally just after its emergence from the superior margin of the intertransverse foramen. It was evident that the pointed stick, six years ago, had with great accuracy torn up the vertebral artery as it lay in its short

horizontal course outside the dura and had put it in communication with the venae comites or some other member of the suboccipital venous plexus. The oversewing succeeded in arresting all bleeding. Recourse to the boiled stethoscope assured us that the fistula had been obliterated by total aneurysmorraphy, a method suited only to relatively small arteries in an inaccessible position. The stethoscope had proved very valuable as a means of leading the dissection to the point of maximum intensity of the bruit. The wound was closed after a four and a half hour operation. Prophylactic systemic penicillin was given for three days. The wound healed without trouble. The bruit remained inaudible at the time of discharge a month later and is absent ten years later.

Case 3 (reported by courtesy of Professor Norman Dott)—A man aged twenty-eight sustained multiple wounds of tibia, lumbar region, wrist, elbow and scapula region by " tommy-gun " bullets in December 1944. He had a wearisome convalescence because of infection. The bullet which entered over the scapula evidently ploughed its way up the neck as high as the atlas, radiographs showing a fracture of the left atlantal and axial transverse processes and the bullet deep to the sigmoid notch of the left mandible. Immediately after wounding he noticed a blowing bruit in his head which worried him more and more as time passed. It seems that there had been a transient contusion of the spinal cord because he had had difficulty in voiding and then in controlling urine at first and also some numbness as high as the lower abdomen, but without paralysis of the leg. His other wounds slowly healed during the next twelve months but head pains, dizziness and insomnia were very troublesome. The patient had been at times difficult to manage; at all events it was some time before the presence of a bruit was definitely observed and the diagnosis of a suboccipital arteriovenous aneurysm made. An excellent surgeon made open angiograms of the left vertebral arterial tree without the step contributing to the verification of the site of the fistulous communication. The left vertebral artery was then ligated at the root of the neck. Two months later in May 1944 the same surgeon made a more direct attack on the aneurysm, ligating the artery in its third portion between the occipital condyle and the transverse process of the atlas. This second operation stopped the bruit but two months later it had returned. A third operation was now undertaken. The jugular vein was divided high and many abnormal vessels were secured; the external carotid artery and its branches were also ligated because it was thought that there might be a contribution from this source. The same high-pitched tinnitus was still present and the bruit, as loud as ever, was audible between the left mastoid and inion, unchanged by cartoid compression. Cerebellar and brain-stem signs were present in the shape of diplopia, unsteady gait and thick speech; there was no deafness; the left corneal reflex was diminished and there was hyperaesthesia of left face. All along he had complained of constant pain deep in the ear. He showed considerable emotional instability, and felt frustrated by his long invalidism from his multiple wounds but particularly because of his head troubles. The patient had now come under the care of Professor Dott and Mr Alexander, who believed that the persistence of the fistula after two ligatures, one of the first portion, the other of the third, must be due to the persistence of communications with muscular and spinal branches. Possibly the ligature of the jugular vein had caused back pressure in the special circumstances of the man's circulation, in that way causing cerebellar signs, but very likely it was the high pressure in the vertebral veins which was the major factor.

Operation (September 1944: Professor Norman Dott and Mr George Alexander)—A horizontal incision was made using the top of the previous suboccipital flap and passing forwards over the base of the mastoid process and then down close under the ear lobe across the tip of the mastoid process and into the neck, where the upper end of the previous cervical incision was used. The suboccipital muscles and the sternomastoid muscle were cut across just below their insertions and the dissection was carried in the subperiosteum along the floor of the skull. The previously made bone defect in the left half of the posterior fossa was encountered; muscles were separated from the dura which was not opened. The sternomastoid was

completely cut through. The accessory nerve was identified and the muscle was slung downwards and forwards, hinging on the accessory nerve. The splenius capitis was also divided close to the bone in its anterior half. The digastric muscle was partly divided but largely retracted. The facial nerve was not actually seen: it lay just in front of the dissection and was retracted forwards with the parotid. This brought the operator down very conveniently on to the transverse process of atlas, which was isolated from its muscular attachments. A pulsating varix could be appreciated just below it. The transverse process was found to be loose, as was the case in Stromeyer's patient (Perrig 1932), having been fractured through its foramen, and after the outer part had been freed from all attachments, it was removed. This exposed the vertebral artery in its foramen, from which it was loosened. Immediately below the foramen and overlying the vertebral artery was a vascular sac about 1 centimetre in diameter, and draining it were three large veins, two descending rather posteriorly and one ascending, winding round in front of the transverse process and appearing on top of the vertebral artery above it. One branch reached inwards immediately above the vertebral artery, and the other forwards towards the jugular foramen, probably joining the jugular vein close to the skull. These venous branches were found to contain arterial blood which pulsated. When they were divided however their distal ends regurgitated only venous blood. The vertebral artery itself was isolated down to the foramen in the second cervical transverse process, and there it was ligated and cut. The fistulous opening thus had been excised with the corresponding segment of the artery. The opening was on the postero-lateral surface of the artery, immediately below the transverse process of atlas, and it was surmounted by the dilated venous sac.

This operation succeeded in its aim. The bruit and pulsations were permanently abolished. The patient, although better in some respects, continued with pain in the hyperaesthetic face. Two further operations were carried out by Mr Alexander, one a fractional, the other a complete division of the fibres of the left trigeminal root, but without entirely ridding the patient of his discomforts. The bullet deep to the sigmoid notch had previously been removed by Mr Alexander. Later still the left cervical sympathetic chain was resected (Professor Dott) because of trophic ulceration of nostril. The patient was well in 1954.

These three cases demonstrate clearly the difficulties that beset the surgeon. They show again that the only certain way of curing these fistulae is by obliterating them. They form a special group because whereas the majority of arteriovenous fistulae occur where a large artery lies beside a large vein there is here no single vein but instead a plexus of veins larger than might be thought. In Figure 6, reprinted here by courtesy of Professor Boileau Grant, the richness of the deep cervical venous plexus is very clearly illustrated. Studying this diagram it is easy to understand how it is that these vertebral arteriovenous fistulae are so singularly formidable surgically. It is impossible to reach the injured vessel without first encountering many of these vessels filled with arterial blood.

PREVIOUSLY REPORTED CASES

The first and magisterial account of injuries and traumatic aneurysms of the vertebral artery was that of Rudolph Matas (1893). In this famous paper he gave an account of the twenty-two injuries (of which nineteen were fatal) and the twenty aneurysms published up to that date. No more than six of the patients with aneurysms had survived, including one patient of his own. Matas made no distinction between saccular and arteriovenous aneurysms, but it is probable that most, if not all, were fistulous. Several of these examples were in the third, the suboccipital, part of the vertebral artery. Korteweg's case (1903) was in this situation—a knife stab injury cured by ligature of the artery between the transverse processes of atlas and axis and forci-pressure on the distal end above the atlas for two days. No comprehensive review of the subject was made for many years until Küttner (1917, 1930) published seven

new cases, two in the situation under review. It may here be remarked that aneurysms of the first part are rarest and of the second part of the vertebral artery rather commoner than those in the third (5; 25; 19—Küttner 1917). The common carotid artery is sometimes injured as well, as in Learmonth's case (1944). Perrig (1932) brought the literature up to date in a review of the forty injuries and sixty-seven aneurysms that he had collected from all sources. In 1941 Ogilvie reported a mycotic arteriovenous aneurysm of the third part of the vertebral artery cured by ligature of the first part. This success runs counter to the consensus of opinion that proximal ligature alone is not only useless but dangerous for arteriovenous aneurysms (Holman 1945). There are evidently exceptions to this rule.

By this time great interest was being aroused by the congenital or arteriosclerotic saccular aneurysms of the vertebral and basilar arteries within the dura mater. These had long been known but had not been thought to be as common as the experience of the last few years has proved (Yaskin and Alpers 1944, Hamby 1952). These aneurysms will not here be further referred to. The commonest congenital arteriovenous malformations seem to be those of the cerebral vessels, and here the question of carotid ligature sometimes arises, giving point to the problem of proximal ligature just mentioned.

Heifetz (1945) had a patient with a vertebral aneurysm at the root of the neck (part one) and in the following year Daniel Elkin and Harris recorded their remarkable experiences with ten cases of vertebral arteriovenous aneurysm among 500 vessel injuries. Three were in the same region as two of the three recorded in the present communication. Elkin (1947) later had three more high in the neck, all of which gave great technical difficulty. More recently Norman, Schmidt and Grow (1950) had an unusual case of congenital arteriovenous fistula in an infant of six months admitted to hospital with cardiac decompensation from the fistulous venous back pressure on the right side of the heart in the way made familiar by the writings of Holman (1937). This patient was cured by excision of the fistula from over the transverse process of the axis and ligatures of the vertebral artery. One of the present writers (R. A. B.) has an excellent adult example of such a case with pronounced changes in the spinal cord and consequent spasticity from similar back pressure, but without cardiac troubles thus far.

Mention must be made of two other individual cases. The first was reported by Makins (1919) in his well known monograph on arterial injuries. There were three cases of damage to the vertebral artery out of 497 cases (0·6 per cent). Makins said: "The third case was operated upon by Colonel Harvey Cushing; the sac was exposed in the suboccipital triangle, and it was found to be impracticable to apply a ligature locally. The sac was plugged, and the artery was secured at the root of the neck. The operation was successful." This is a somewhat colourless account of what may well have been a dramatic intervention. The other case is one referred to by Penfield (1946) in discussion on Elkin's series. Penfield had had an intraspinal but extradural congenital arteriovenous malformation in which the proximal end of the artery was tied between C.5 and 6 transverse processes and the distal intradurally alongside the medulla oblongata. It is improbable that this operation would be feasible or successful in the traumatic extraspinal types because the cerebellar approach would have to be made in part through the aneurysmal field with its formidable complex of vessels. Once the surgeon has really got into the radicles of these fistulae and the wound is full of blood it is no easy matter under duress to open the posterior fossa and secure the vessel there neatly. It may be that this Penfield procedure is a correct treatment, and it would naturally make a special appeal to those familiar with intradural procedures, namely the neurosurgeons. At the worst, if the bruit was not completely abolished by the double ligature, it should be an easier and less anxious matter to make a direct attack at a second operation on the congeries of vessels that make up these aneurysms. But we must point to a difference in aim in arterial ligatures for the vertebral traumatic and congenital types. In the former the object is to arrest the circulation totally. In the latter, if the spinal cord is involved, a reduction in circulation and not complete standstill is all that is permissible.

CHOICE OF OPERATION

Some tentative suggestions have just been made on this subject. The site of election for ligature of the vertebral artery is undoubtedly in that part of its course where it passes between its foramina in the transverse processes of the axis and atlas. The primary approach should be postero-lateral. The anatomy of the approach to the vessel here has been worked out in the diagrams illustrating this communication from dissections by one of us (R. A. B.) and drawn by Miss Dorothy Davison. The vessel was ligatured here in all three of the present cases. If the diagrams have a defect it is that they show the operation field rather too much as if it were primarily posterior. It should be noted that it is one thing to print diagrams of the approach to a normal vessel but quite another to dissect the vessel out in the presence of an arteriovenous fistula. The reason is that the exposure of the aneurysm requires the carrying posteriorly of the incision towards the occiput after the division of the sternomastoid and the muscles deeper to it. In doing this it is the rule rather than the exception for the operator to open into some of the vessels draining away arterial blood from the aneurysm. As soon as this happens his careful plans for the neat surgery of the lesion vanish and all his wits and judgment are required to keep the bleeding under reasonable control while the artery is dissected out and tied. As soon as one source of bleeding is stilled by forci-pressure or diathermy the aneurysm seems to bleed worse from somewhere else. Packing or finger pressure will have to be resorted to and either of these controls is likely to get in the way of dissection of the artery. For that reason it is wise to ensure the exposure of the commencement of the third part of the artery at an early stage before, as far as possible, the aneurysm has been disturbed. In Case 2 this precaution was not observed, making the operation more difficult than it should have been. It must be noted that these arteriovenous aneurysms are curious things in that they are almost like extremely vascular sponges infiltrating, as it seems, the muscles of the suboccipital region. They overlie the parent artery which is the last thing to be reached. Nor are the surgeon's troubles over when the proximal ligature has been applied. The vertebral artery not only gives off numerous small muscular branches, but there is also the remarkable potential back flow from its fellow of the opposite side through the confluence of the two vessels in the formation of the basilar artery. In Case 2 the artery in the whole of its extradural third part was unrecognisable save for a short cuff just as it penetrated the occipito-atlantal ligament. A similar difficulty in identifying the vessel as a trunk was clearly present in many cases reported by others.

The sterilised stethoscope used in Cases 1 and 2 proved to be of the greatest service in deciding whether or not the fistulae had been obliterated. More than once in the course of the operation it seemed as if success had been achieved until correction was given by the stethoscope. This instrument was useful in another way, in disclosing where the maximal bruit was. That is helpful.

It is remarkable that an artery the size of the vertebral can give rise to such ferocious bleeding. It certainly does. If it were decided to tie the distal and proximal ligature as in Penfield's case the surgeon would have to restrain his enthusiasm for a wide exposure and avoid the aneurysmal region not only laterally but in the exposure of the occipital bone preparatory to opening the skull. We can foresee the greatest difficulties. Penfield kept out of trouble by tying the vessel at C.5–6 level. Elkin and Harris thought it not much help to ligate the vertebral artery far away from the aneurysm—say at the classical site in the stretch between the subclavian and the transverse process of the sixth cervical vertebra. We are sure that this is correct. It was not successful in Case 3. The artery has so many anastomotic channels above that level in its upward course that ligature at best reduces the blood flow, it does not arrest it. Hence ligature between the atlas and axis is more certain to succeed. It must be noted, however, that low proximal ligatures with packing of the bleeding aneurysmal complex with gauze have succeeded or seem to have succeeded in some cases, such as Cushing's.

One useful comment must now be added. The picture that the surgeon carries in his mind of the course and appearance of the artery is sure to be that of the diagrams in books or of museum specimens. It was clear to us in the course of our operations that the rotation of the head necessary for exposure caused a considerable change in the artery's uppermost intertransverse and suboccipital course. The artery is always figured as straight and taut here between the processes but that cannot be so; there must be some slack to allow of strong rotation of the head. In Figures 4 and 5 the differences are illustrated from dissections by R. A. Bailey who has particularly studied this point. The changes are great enough to make the surgeon wonder whether the vessel he sees really is the vertebral artery. The swing of the atlas acts like an outrigger, carrying the vertebral artery forwards so that the parts of the vessel above and below the transverse process of the atlas lie almost parallel to one another.

NEUROLOGICAL EFFECTS OF VERTEBRAL ARTERIOVENOUS ANEURYSM

Neurological signs were present in only two cases of our group, Case 1 and Professor Dott's case (Case 3). The history in this latter case directly after wounding showed that the upper end of the spinal cord had been contused. But later other neurological troubles developed of a cerebellar or ponto-medullary kind that must have been caused by vicious retrograde circulation. It would be supposed that the most potent cause of neurological damage would be the initial trauma, as in our Case 1 where the missile had wounded the third division of the trigeminal nerve (see Jefferson and Schorstein 1955 on trigeminal injuries) in its passage through the face and then the jugular group of nerves and the cervical sympathetic as it swept past the skull base. Coincidental injuries to the upper trunks of the brachial plexus have been reported in injuries at lower levels. Elkin and Harris had such cases as well as those with facial and auditory lesions and an injury to the thoracic duct.

A noteworthy feature is that not only our own example of a stab wound, but, as perusal of the literature discloses, the majority of those observed by others have been in the left side of the neck. This is a natural lateralisation for wounding by a right-handed assailant, attacking from the front or side. The right-sided stabs have usually been inflicted from behind.

We shall not comment further because the neurological signs presented by the initial trauma do not come properly within the scope of this paper. Those consequent either on the aneurysm or on its treatment are more to the point. And here it can be said that it is an impressive fact that ligature of one vertebral artery does so little harm. The reason is not far to seek—its very free whole-trunk confluence with its fellow of the opposite side.

The only fatalities likely would be in the rare instances where the opposite artery is no more than vestigial, a very rare event. We recall that Alexander of Liverpool (1889) tied the vertebral arteries, often both at one sitting, thirty-six times with no more than three deaths, for epilepsy in the mistaken belief that epileptic fits started from the medulla. On the other hand, Dandy's (1944) single patient treated by bilateral ligature died instantly when the second vessel was tied some weeks after the first. No case of fatal ischaemia of the brain stem has yet been reported in subjects operated upon for vertebral arteriovenous aneurysm. The causes of death were tabulated by Matas as haemorrhage, shock, sepsis and exhaustion and lastly cerebral complications. They remain the same. It must be noted as to sepsis and exhaustion that some aneurysms were due to cervical caries and all the deaths from infection occurred in years gone by. Neurological signs might arise secondarily from high venous back pressure in the intradural veins because of the very free communications that exist through the intervertebral foramina between the venous systems of the nuchal muscles and the intraspinal veins. It is likely that the increase in neurological disturbances that gradually took place in Case 3 was due to this cause—a slow build up of pressure, in fact a reversed circulation, in the blood supply of the upper spinal cord, cerebellum and other structures in the posterior fossa. Apparently none of the patients observed by Elkin and Harris suffered

in this way. But it has to be admitted that almost all the published examples have been in the care of general surgeons who, however distinguished otherwise, were not noteworthy neurological experts. It is consequently difficult to distinguish in their accounts of the clinical status of their patients the immediate from the late effects. It should be said that in our own Case 2, in which an arteriovenous fistula had existed for six years, no neurological signs developed during the intervening period. Evidently disturbances in, or reversal of the, venous circulation does not necessarily lead to recognisable dysfunction. Professor Dott finally attributed much of the neurological disturbance in Case 3 to temporary drug addiction.

THE ROLE OF ANGIOGRAPHY

In the older literature there has been the greatest confusion as to the identity of the vessel at fault. It is natural that the carotids should have been the first choice and should have been in the past so often ligatured before the aneurysmal source was recognised. This mistake was made in over half the cases, though not in recent years.

A source of difficulty has been that in many cases, as in Case 1, the pulsation and bruit could be reduced by carotid compression. But, as Fraeys (1848) pointed out and proved by cadaver experiments, pressure about the level of Chassaignac's tubercle can occlude both the carotid artery and the first part of the vertebral. There is, then, here a possible source of false deduction.

The occipital artery was often the next choice after the carotid, but this vessel does not in fact readily lend itself to a fistulous type of lesion, being too closely invested in muscle planes. The vertebral artery's case is different because it has a venous plexus round it and it is protected from close muscular investment whether in its second or its third part. Muscles certainly cover it, as must be the case with any vessel as deeply situated as this, but a tunnel is left for it, either in its interrupted bony canal or as it lies on the atlas. It is by contrast improbable that an occipital artery arteriovenous fistula has ever occurred.

At the present day angiography should prove an easy way of differentiation, either by exclusion on carotid angiography or by the positive proof of vertebral angiography. It was used in cases 2 and 3 here inconclusively but at a date when the techniques were not so good nor the results so certain as they are today. It is of interest to add that the high route for angiography by the mastoid tip, puncturing the vertebral artery as it winds round the back of the lateral mass of the atlas, has itself resulted in two or three examples (unpublished) in which a bruit has developed. The presumption is that the puncture has produced a leak into the vertebral venous plexus, but in no case has it been more than a minor inconvenience. The fact, nevertheless, adds point to the remarks made above of the relative nakedness of the vessel and its proneness on anatomical grounds to fistula should precisely the right lesion be inflicted upon it.

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THE HAND IN SURGERY *

NORMAN CAPENER, EXETER, ENGLAND

> The old farm labourer moved away " in the enjoyment of his theory that every tool has two ends, one working on the material, the other on the man."
>
> John Halsham: *Lonewood Corner* (1907)

For a surgeon more than for most people the hand is one part of the body whose function is constantly under observation. Until the pioneer work of Kanavel and Bunnell, familiarity has, even amongst surgeons, tended to breed neglect. We in Great Britain owe much to that delightful treatise upon " the principles of anatomy as seen in the hand " by the late Frederic Wood Jones. A surgeon's study of his own hands should give him better appreciation of

1—" Study of Surgeon's Hands 1947." A drawing by the sculptor Barbara Hepworth which illustrates many points dealt with in this paper. (Reproduced by permission of the artist.)

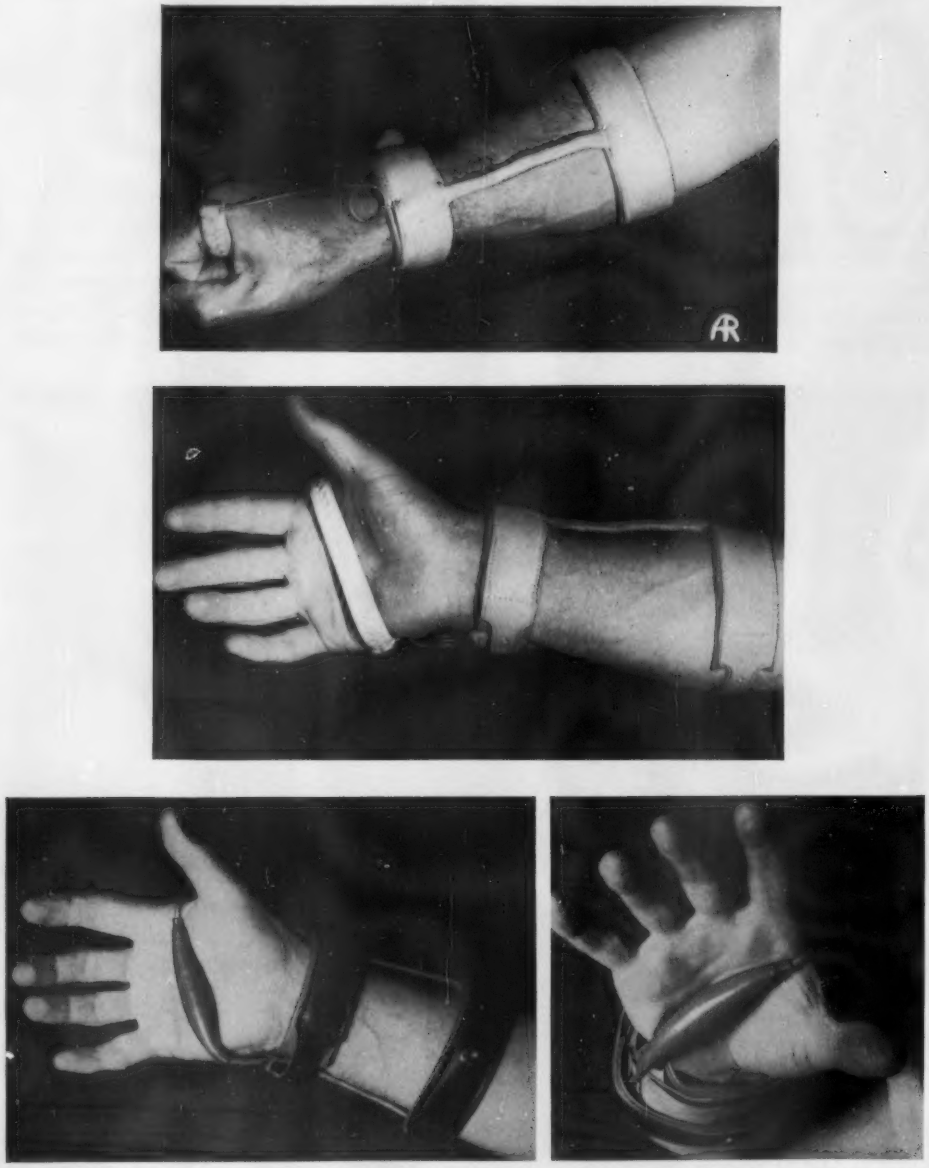
the manual disabilities of other craftsmen. That is the intention of this paper. The subject is complex, for apart from the anatomical and physiological facts of motor function, hands in action reflect so much of intellect and character and furthermore are instruments of sensibility and of expression. My discussion must therefore be selective and somewhat superficial.

* Revised from the previously unpublished presidential address given to the Orthopaedic Section of the Royal Society of Medicine, March 7, 1950.

2—Anatomical and physiological positions in lateral radiographs with a tangential view of the palmar concavity at the metacarpal heads.

THE ORCHESTRATION OF THE HAND

One of my particular interests in the hand has been to work with my friend, the late Mr F. W. Suter, upon improvements in the design of splints for physiological control. We



3—Cock-up splints which allow maximal palmar sensory function and are shaped with due regard to the double palmar obliquity.

were concerned to develop the idea of mobility, or "liveliness," which also has been the interest of other workers. We were guided by the idea that, in the hand, function involves an orchestration of motor and sensory activities. Absence of, or defect in, any of the instruments

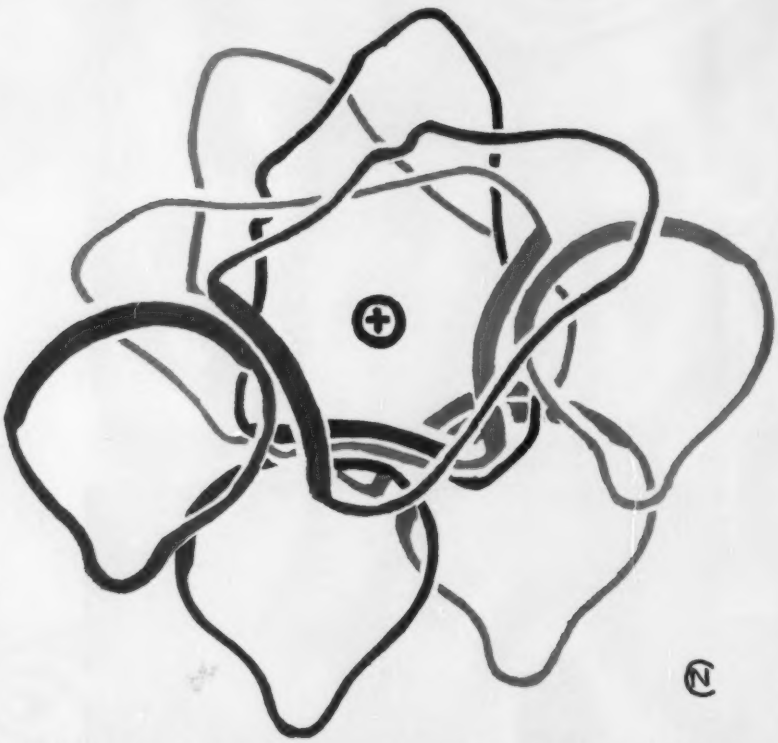
of this orchestra, produces imbalance or discord in the rest. Substitution for lost instruments may restore balance because of the harmonics of reflex activity induced by sensory stimuli, especially those of proprioceptive nature. Such artificial balance may aid the restoration of

4—The dynamic tripod illustrated by the use of scissors and screwdriver, and also well seen in operative view of scalpel and forceps. The latter also indicates the common position of relative pronation and adduction.

natural balance if other circumstances permit it. In a musical orchestra the disposition of its members one to another and in relation to the conductor is important for well tempered effects. An essential instrument of the hand is its palmar skin surface which must be open and used (Figs. 2 and 3).

MANUAL POSTURE

The disposition of the hand is important for, with a member (or group of members) having such infinite variables of purpose, the surgeon may be committed to the selection of one position either temporarily or permanently, in the treatment of disease or injury. We all recognise the dreadful notion provided by the "anatomical position," the use of which is so rare in life and is so unphysiological for therapeutic purposes. For the majority of purposes the hand basically assumes flexion to variable extent of all the phalanges with some opposition of the thumb. At the wrist a little ulnar deviation with the forearm pronated is very much commoner than is usually appreciated. Movements from this more or less resting position



5—Sectional diagram of the radius and ulna in four phases of pronation-supination. Red = supination, green = semi-supination, black = mid pronation, blue = maximal pronation. A central axis of rotation is marked +, but the axis is variable.

are, of course, infinitely complex. Nevertheless, one is impressed by the great use made of the flexed, adducted and pronated position of the wrist. Doubtless this is related to the frequent use of the hands upon material below their level and in front of the mid-line of the body. In considering the palmar concavity, the transverse obliquity of the metacarpal flexion crease is well recognised, but insufficient notice is taken of the obliquity of the second to the fifth metacarpal heads in the antero-posterior plane. When the thumb and fifth metacarpal bones continue the line of the forearm bones the second metacarpal is seen to take the most extended position. Objects, such as a fishing rod or a mallet, gripped in the palm thus take up an oblique position in two planes, that is, both antero-posterior and transverse. These are features of importance in the design of physiological splints.

THE HAND IN ACTION

In the hand are found many aspects of duality; static and dynamic, motor and sensory. One is seen in a differentiation of the ulnar and radial portions. The ring and little fingers are used largely for support and static control while the thumb, index and middle fingers form

6—Radiographs of the elbow joint in the three phases of pronation (A), mid-position (B) and supination (C). At (B) the ulna is relatively extended. (D) Double exposure of forearm in pronation and supination.

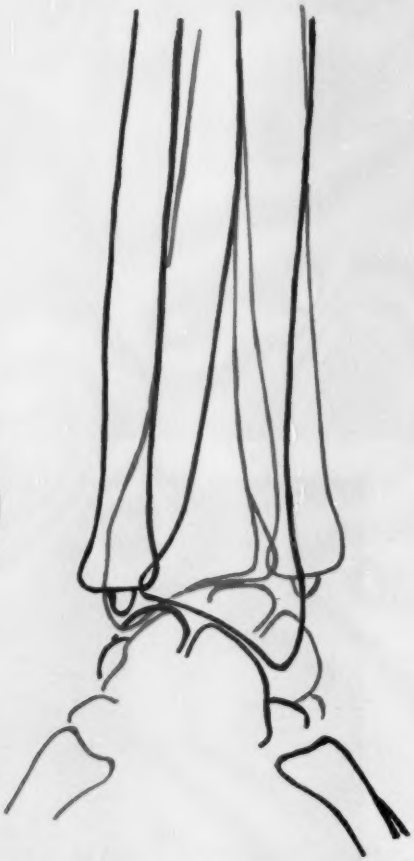
a *dynamic tripod.* The division is not clear cut; each element depends on the other and the middle finger takes a share of both activities (Fig. 4).

There are three major functional axes: longitudinal, antero-posterior and transverse. As in most biological articulations, movements are not around fixed axes. In longitudinal rotation (pronation—supination) it is difficult to accept the common view that the radius rotates around the lower end of the ulna, which remains a fixed point. Most anatomists*

* Recently in 1954 R. J. Last has given an admirable presentation of this matter.

recognise this but many surgeons, including even Bunnell, are not clear upon the point. If the radius only rotated around the ulna, the dominance of what I have called the dynamic tripod would be less, and the use of rotating instruments, such as a screwdriver, would be difficult. Observation of one's own hand confirmed by radiography shows that during prone-supination, the lower end of the ulna usually describes an arc from side to side and in a plane posterior to the radius, while the latter rotates in an opposite direction. Figures 5 to 8 demonstrate the movements schematically and radiologically. I conclude that the axis of longitudinal

7—Superimposed tracings of the three radiographs, A, B and C, from the previous illustration. Pronation = blue, mid-position = black, supination = red.



8—Tracing of a double exposure of forearm in pronation and supination here indicating an axis almost centrally in the lower end of the radius.

rotation of the hand and forearm is generally to be found within the centre of the radius at its lower end, but it may be anywhere between the radial and ulnar styloid processes. Artery forceps, scissors and screwdrivers demonstrate the necessity for this. With a tool such as the screwdriver, the mechanical axis may be affected by the point of " peripheral fixation." I mention this now because we tend to forget that tools are not simply carried in the hand; they work upon material by contact, and, in the reverse direction, the material influences the limb which holds the tools.

Accepting that some ulnar deviation of the hand is part of its physiological mean, further medial and lateral movement occurs at both the radio-carpal and mid-carpal joints (Fig. 9). For this the approximate axis lies near the centre of the ulnar border of the capitate bone. Rarely, however, is such movement a simple matter of abduction and adduction. It has been noted by Bunnell that lateral deviation movements are most easily carried out radialwards with extension, and ulnarwards with flexion. In these two directions lies the plane of physiological lateral movement, and it is significant that it corresponds respectively with the action of the radial carpal extensors and the flexor carpi ulnaris (Fig. 10). It also corresponds strikingly with the obliquity already described in the physiologically poised hand, due to the unequal extension of the second to the fifth metacarpal bones. This oblique transverse movement is

9—Carpal abduction and adduction. Radiographic studies of the anatomical range of movement which is more than that of the wrist joint.

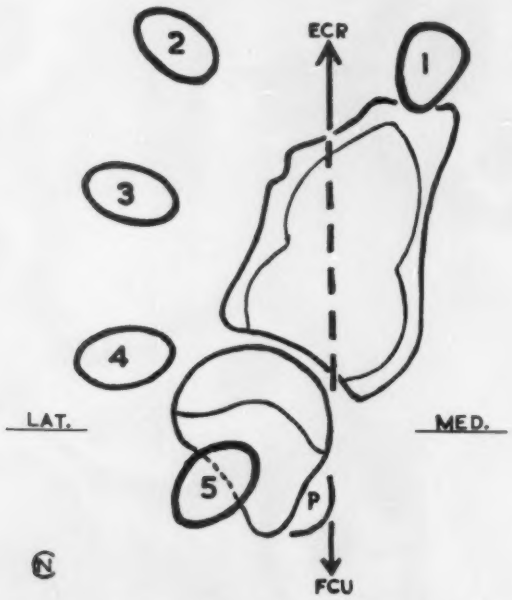
well seen in the use of a mallet. Furthermore, it will be noticed that in the lateral movement of radial deviation and extension, there is slight pronation of the forearm: conversely, in medial movement transversely, with ulnar deviation and flexion there is slight supination. This supination is partly due to the action of the biceps and is a synergic braking action which limits extension of the elbow at the same time. This is one type of paradoxical muscular action or lengthening contraction. It is important in all manual occupations involving a swinging action of the forearm with extension of the elbow joint and strong action of the wrist in an ulnar direction. It is thus seen in the use of most tools of percussion as, for example, by sculptors, carpenters, stonemasons, motor mechanics, fly fishermen, tennis players and orthopaedic surgeons.

The third major action of the hand unit, namely the antero-posterior sliding rotatory movement around the transverse axes of the radio-carpal and mid-carpal joints, is like the previously mentioned lateral movement, rarely one of simple flexion and extension,

for with flexion there is usually some ulnar adduction and with extension some radial abduction.

THE CONTROL OF INSTRUMENTS

Instruments can be differentiated into two main groups: those in which the holding is mostly digital, and those which are clenched in the palm. Both groups can also be divided according to their direction; that is whether towards the ulnar or radial sides of the hand. The latter has an interesting functional significance, comparable to the difference between low and high mechanical gears. In holding a mallet, with the fingers tightly wound around its handle, the position is one of obliquity, much greater than the metacarpal obliquity already

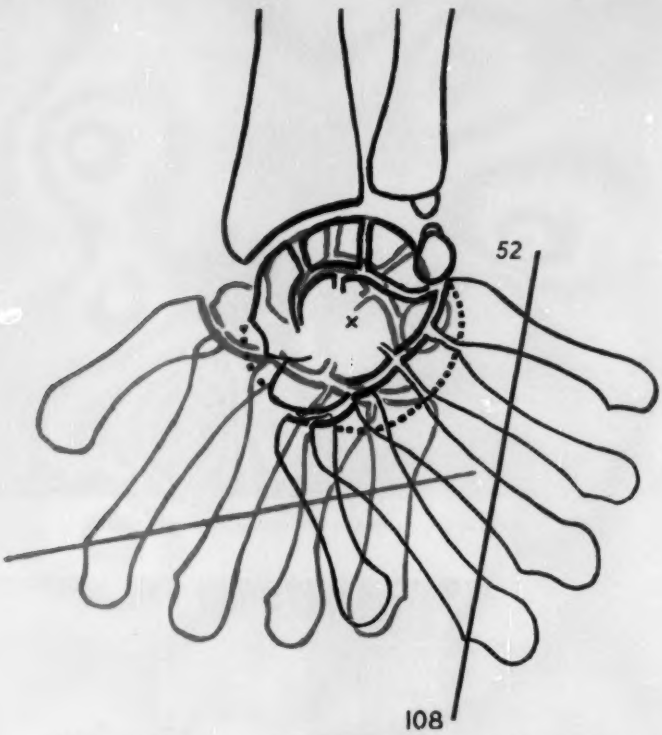


10—Scheme of the lower end of the radius and ulna in relation to the metacarpals marked 1 to 5. The wrist is somewhat pronated. "Medial" and "lateral" refer to the ulna. ECR = line of action of extensors carpi radialis. FCU = line of pull of flexor carpi ulnaris. Between them is the oblique line of grip and the usual plane of physiological "medio-lateral" movement. P = pisiform.

mentioned. The active end of the tool and the centre of gravity is placed beyond the radial border of the hand. Dynamic control is given by the thumb, index and middle fingers, which press the butt of the tool against what a craftsman calls the "heel of the hand" which is the hypothenar prominence and the pisiform bone. The amount of the obliquity of this grip is 35 degrees or more to the transverse. These features are represented in the diagram (Fig. 11), in which a circle is marked showing a continuation of the curvature of the radio-carpal articular surfaces. Diametrically opposite the radius the circle is near the normal oblique line of grip at the heel of the palm. If the hand is moved in a counter-clockwise direction around the circle it will be seen that for 20 degrees of movement the ulnar end of a gripped handle moves in the ratio of about 1 to 2 in comparison with the radial end. Thus a form of gearing is given, the effect of which is to enlarge the range of movement of tools pointing to the radial side. The longer the tool and the more obliquely it is held, the greater the magnification of

the range of excursion of its tip, and of its momentum, features well seen when casting with a fishing rod. Conversely for tools pointing ulnarwards, for example a chisel, the excursion of the tip is reduced but power (at the wrist) is increased. The muscle which is the " tendo achillis " of the hand is the flexor carpi ulnaris, and it will be noted that its point of attachment to the pisiform bone lies close to the circumference of the circle of lateral deviation movement.

The screwdriver, bone-cutting forceps and bone nibblers are, like the mallet, tools which are clenched in the palm and directed in a radial direction, though in these instruments the

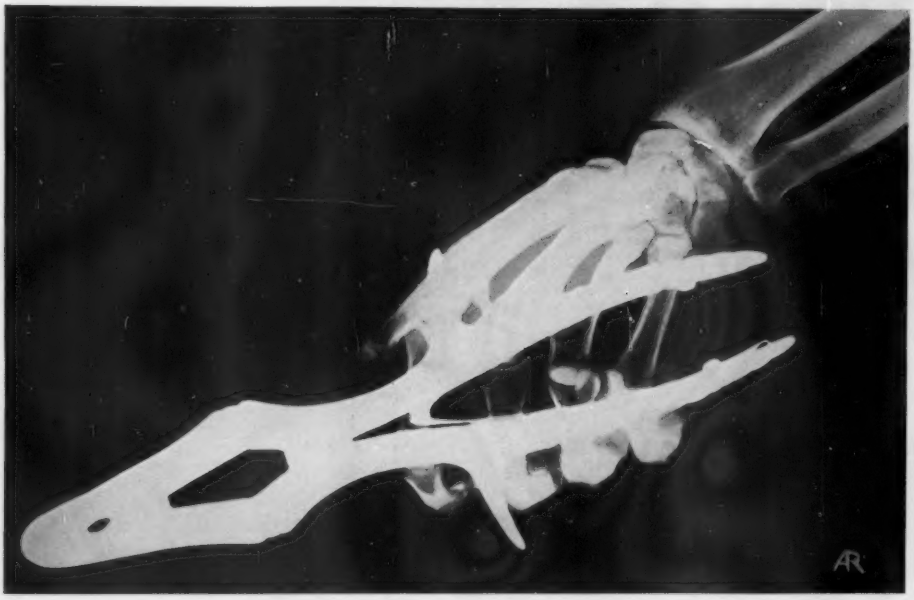


11—Lateral movement and the transverse angle of grip. Overlapping tracings of the extremes from the previous illustration. The straight line marks the position of a gripped handle showing the gearing effect described in the text. The figures give the distance in millimetres moved by the two ends of a handle from full abduction (red) to full adduction (black).

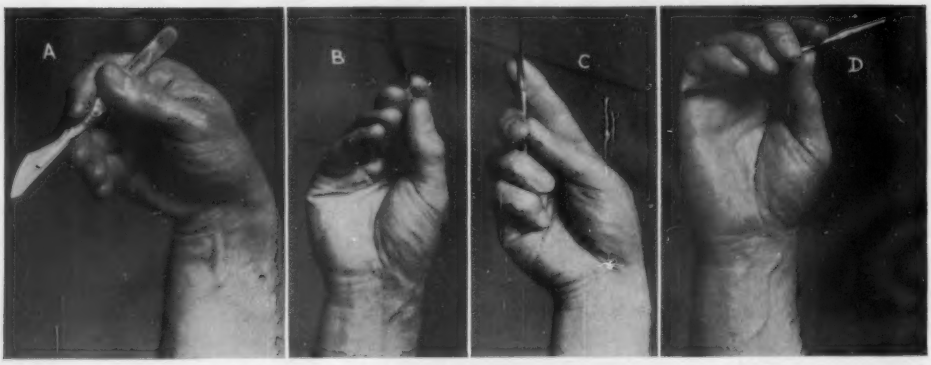
obliquity is much greater and nearly approaches the vertical axis of the limb. For them the heel-like function is taken in part by the thenar muscles (Fig. 12).

Of tools that are digitally operated, scissors and haemostats are good examples. The former is the ancestor of all surgical instruments which have double-ringed handles with two centrally hinged blades. Almost invariably these instruments are directed towards or beyond the radial border of the hand. The simplest hold is by the elements of the dynamic tripod, direction being given by the index finger, force applied by the thumb against the more passive support of the middle finger. For added support and control, we generally add the ring finger. The basic alignment of such instruments is in a plane considerably anterior to, but parallel with, the longitudinal axis of the forearm. The muscular control is complex. To obtain

proper shearing effect with the blades of scissors and, in haemostats, to obtain freedom of sliding of the locking handle notches, there is added (to the simple compressive action between the thumb and middle finger) a forward or abduction movement of the thumb, and a flexion movement of the distal phalanges of the middle finger. For the haemostat, this action is



12—Extreme angle of grip as seen in the holding of a bone nibbler.



13—The control of the scalpel. A and B the ulnar-directed dissecting position. C and D the radially directed position for power. From A to D is seen the almost complete sphere of action in one plane representing approximately 180 degrees.

greatly emphasised in the process of unlocking it. The compressive effect of the thumb is applied with the interphalangeal joint in extension, and this is brought about largely by the action of the flexor pollicis brevis and the opponens muscles acting by flexion of the proximal phalanx.

In the use of the scalpel, both digital and palmar holding play parts of variable importance

(Fig. 13). It is not easy to analyse the action involved, as is to be expected of an instrument which so closely resembles the pen in its integration with mind. The scalpel is used either in a radial or ulnar direction, in different manner and for different purposes. In the first direction the butt of the scalpel rests in the palm or along the ulnar border of the hand, and the angle of the instrument is controlled by the ulnar fingers. Its blade and point are directed beyond the radial border and force is imparted through the dynamic tripod. This is the position for the longer and heavier strokes of incisions. In the second position, that used for fine dissection, the knife is balanced in front of its centre of gravity on the apex of the dynamic tripod, with the butt resting on the fleshy part of the first dorsal interosseous space. This method of holding ensures that the weight of the scalpel rests upon the hand and away from the material; thus it resembles more closely the pen position and makes for lightness of touch

14—The scalpel and dissecting forceps in the mid (visual) line dissecting position—both ulnar directed. Left hand pronated; right hand in mid-position. A particularly macular visual activity with hands in separate halves of visual field (cf. mallet and chisel).

between the instrument tip and point of contact. Thus held the scalpel is directed beyond the ulnar border of the hand.

Dissecting forceps is held directed towards the ulnar border of the hand with the centre of gravity at the apex of the dynamic tripod (Fig. 14). It forms a compressible spring, with differences between one pair and another in both temper and length. Too strong a spring causes fatigue and inefficiency. Even in Ogilvie's well balanced instrument, the varying power used for special purposes is remarkable. Pressing the tips together may require about 1½ lb. of force, but if the complete holding surfaces are to be approximated pressures up to 8 lb. may be necessary—a tiring business for those who use a no-touch technique. The effect of peripheral fixation is extremely well seen in the use of this instrument, for the force at the tip of the digit is exerted in extension, which in this case is a function of the flexor of the proximal joint (flexor sublimis in the finger and flexor pollicis brevis in the thumb). Dissecting forceps in use resemble the dissecting position of the knife, not only in being directed ulnarwards, but

in having a sphere of action which is towards the mid-line of the operating field. It is the more passive tool of these two instruments when used in partnership.

The dissecting forceps offers a pretty example of von Baeyer's principle of the closed kinetic chain and the reversal of muscular action by peripheral fixation: a feature which Sir Harry Platt has emphasised as important in manual workers and as a causal factor in many deformities (Fig. 15).

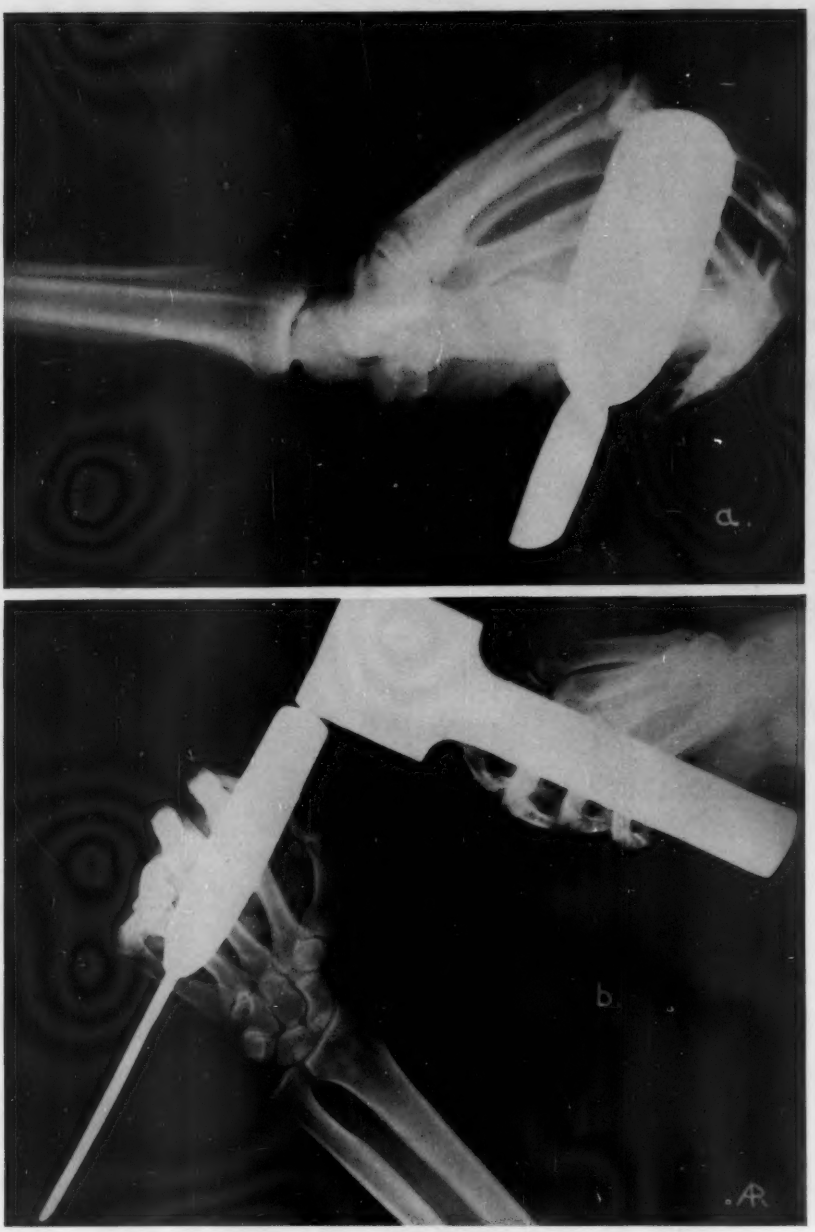
The chisel, representative of many similar tools, is palmar clenched and its position resembles that of the mallet, excepting that the active end points beyond the ulnar border and generally away from the mid-line of operation. The obliquity of the palmar grip tends naturally to direct the blade proximally from the radial to the ulnar side and the handle is

15—The digital hold of Ogilvie's dissecting forceps. An interesting demonstration of von Baeyer's principle of the " closed kinetic chain " and the alteration of muscular action by peripheral fixation: here in its simplest form, the flexor sublimis as an extensor of the distal phalanx.

placed more dorsally than the cutting edge. Because of this, if the cutting edge has to be directed forwards, the hand has to be brought into increased radial deviation and extension. The partnership of the chisel with the right-handed mallet is characteristic of many other bimanual operations of the craftsman. A special type of skill is needed for these left-handed tools which is at least as important as the skill of the right hand, and deserves special consideration (Fig. 16).

HANDEDNESS

What I have said leads me naturally to consider handedness and dexterity. Focillon, a French writer on aesthetics, wrote: " The hands are not a pair of passively identical twins. Nor are they to be distinguished like two children of unequal talent; one trained in all skills,

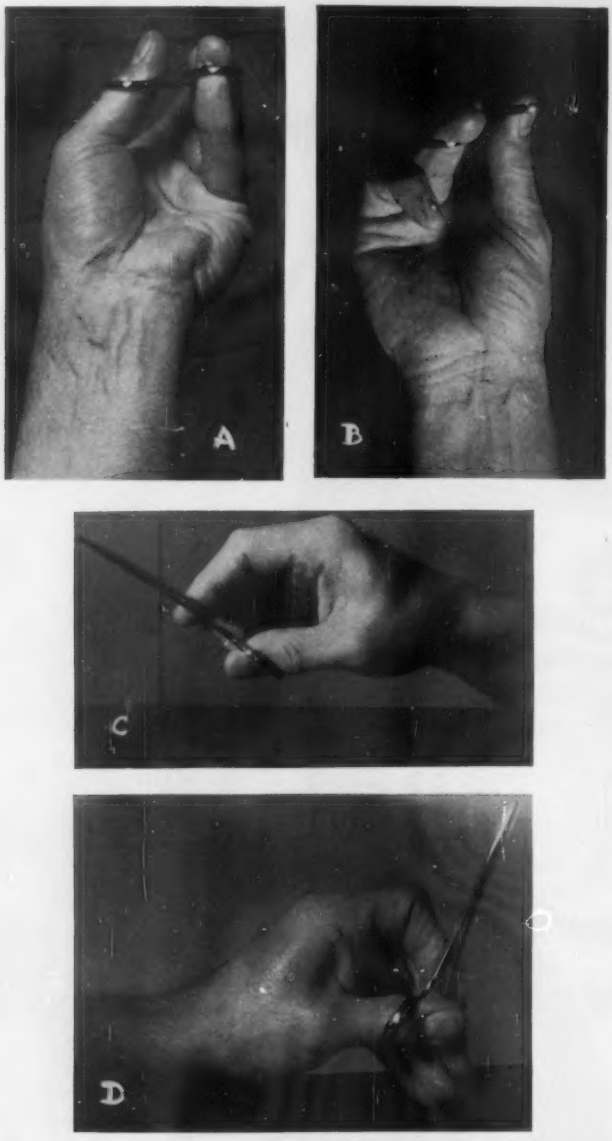


16—The chisel and mallet. Radiographs: *a*) showing the double obliquity of grip of the ulnar-directed chisel; *b*) showing the dynamic of the mallet against the statically controlled left hand held chisel.

the other a serf. I do not believe in the eminent dignity of the right hand. Deprived of the left it retires into a painful almost sterile solitude."

That the left hand can be made to perform all the duties of the right we know in surgery,

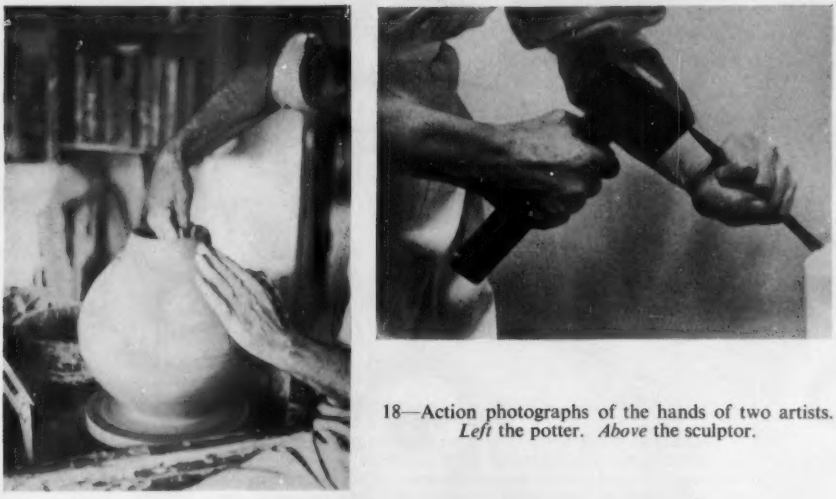
but for efficiency we delegate some functions to the right and others to the left. This is the same for all crafts.



17—The action grip of scissors or unclipping action for haemostat forceps. A and D = left hand; B and C = right hand. The separation of the handles at right angles to the plane of action is the same in both hands but performed by entirely different joint movements and muscles.

Some of our instruments can be used in either hand and the control in one is the mirror of that in the other, and the muscles used are the same. As examples are the scalpel, dissecting forceps, mallet and chisel, though each is generally used for preference in either right or left

hand only. Other instruments which are peculiarly right handed in design require an entirely different action when used in the left. Of these the scissors and haemostat are typical and the first ones which junior assistants find difficult to control left-handedly. The actual movements involved in the use of right-handed scissors in the left should be noted. With the compressive action between the thumb and middle or ring fingers, it is necessary to add extension (instead of flexor) action to the terminal phalanx of the finger, and adduction (instead of abduction) to the thumb. There is a reversal of pressure upon the rings of the instrument with a consequent tendency to twist it so that the shearing surfaces lie in the vertical rather than the horizontal plane required for cutting most ligatures. In the use of this instrument by the surgeon as a dissecting instrument in the left hand, the difficulties are not apparent. Few operating theatres possess left-handed scissors: but I think they have a place in the hand of a novice assistant. Haemostats in the left hand present similar difficulties especially with the force required to undo the clips. All sorts of tricks are employed to overcome what is a relatively slight inconvenience to an experienced surgeon. It certainly is not necessary to have left-handed haemostats (Fig. 17).



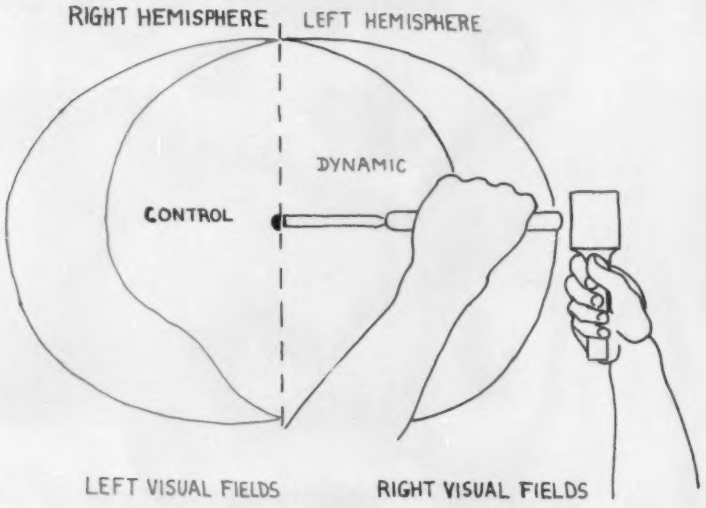
18—Action photographs of the hands of two artists. *Left* the potter. *Above* the sculptor.

As with mallet and chisel so with the tying of ligatures we have an excellent example of bimanual partnership in surgery. Surgeons who do this with their fingers develop in each hand a high degree of skill, and whether the right or the left is the more active or passive, for most of us it is not easy to transfer the actions.

This special skill in partnership between two hands on separate tools is also seen where both hands are used on one tool. Each hand then retains the functional characteristics which it has when working separately. For dynamic power and decision, the right hand is used; where passive support and precise control are required the left hand is preferred. When these two characteristics are brought to perfection, we have two of the essentials of craftsmanship. The difference between them is, I think, that of one we have a greater consciousness. Perhaps it is for this reason that many people unjustly place the left hand in a lower category. It is almost as if we could attribute to the right hand masculine, and to the left hand feminine characteristics. I would even regard the radial and ulnar halves of the hand as having a similar differentiation of character. It may well be that one of the problems of the modern world is to emancipate the left hand.

As an example of craftsmanship at its best, I present a photograph which I took of an artist in pottery (Fig. 18). The wheel is spinning clockwise at high speed as the potter moulds the outside with the left hand while the right shapes the inner concavity. Although this is not the usual English way of throwing a pot it does rather peculiarly illustrate my simile of male and female characteristics. It certainly is an excellent example of bimanual skill such as is also well seen in the playing of a violinist, or in the work of a sculptor, as I also illustrate.

Such differentiated manual skill is probably a more desirable thing than ambidexterity, which has been defined as " the ability to use both hands equally badly." True ambidexterity is a rarity though there have been noteworthy examples amongst surgeons, but it is not necessarily an achievement of which to be proud. A form of ambidexterity is relatively common amongst mental defectives. It is really ambisinistrality due to a lesion of the major cerebral hemisphere.



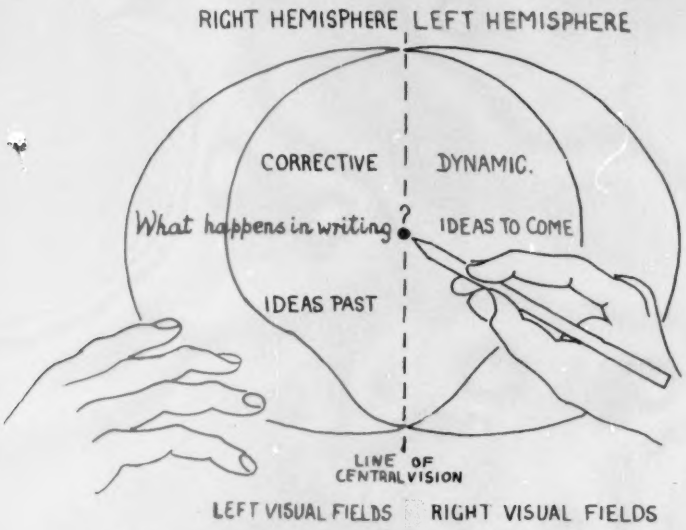
19—Visual field and cerebral dominance in relation to bimanual activity. Diagrammatically the visual fields of both eyes are represented. Macular vision concentrated upon chisel edge. Both hands lie within the scope of the right visual fields (and under the visual control of the left cerebral hemisphere). The control of depth and direction is within the scope of the left visual fields and right cerebral hemisphere.

VISION AND THE HAND

In my original presentation of this paper I discussed handedness in relation to visual and cerebral hemisphere dominance. Without venturing too far into speculation I must repeat a few observations here. Lord Adrian has drawn attention to the fact that the eyes have dictated the general arrangement of the cerebral hemispheres and so of the crossed spinal tracts through the camera structure which focuses objects in the left of space upon the right half of each retina and vice versa. As the eye is therefore not represented as an entity hemispherically neurologists discount the question of eye dominance though many people constantly aim or " point " with one or other eye. The latter, however, is not a reliable guide to handedness or to cerebral hemisphere dominance. I have wondered whether there were not another form of visual dominance—a visual field dominance of left or right: for there are some interesting differentiating features to be noticed in our use of tools. Consider first the use of a mallet and chisel (Fig. 19). The latter held in the left hand is usually directed towards the left away from the mid-line of the body. When it is so held macular vision is concentrated upon

the cutting edge. What lies beyond and to the left is perceived, through the right homonomous fields of vision and by the right occipital cortex, that is of the " minor " hemisphere. The chisel handle and the point of impact by the mallet are not seen by macular vision, but are received through the left ocular fields, by the left or " major " cerebral hemisphere. There is thus particular association of the left visual cortex with the left parietal cortex in the dynamics of the mallet, while the right cerebral hemisphere controls the direction and fine adjustments of more static type. Similar differentiated use of the left visual fields occur in the use of many tools and of other implements in sport, *e.g.*, cricket, golf.

What happens in writing and reading (Fig. 20)? In applying pen to paper and proceeding from left to right, the two maculae are concentrated upon the pen point. What lies to the right, namely what we may call "the ideas to come" or dynamics, is a concern of the left cerebrum, in which we perceive the words before they are formed. What lies to the left of the point, that is, " ideas past " is received through the right visual field by the minor right hemisphere ("the



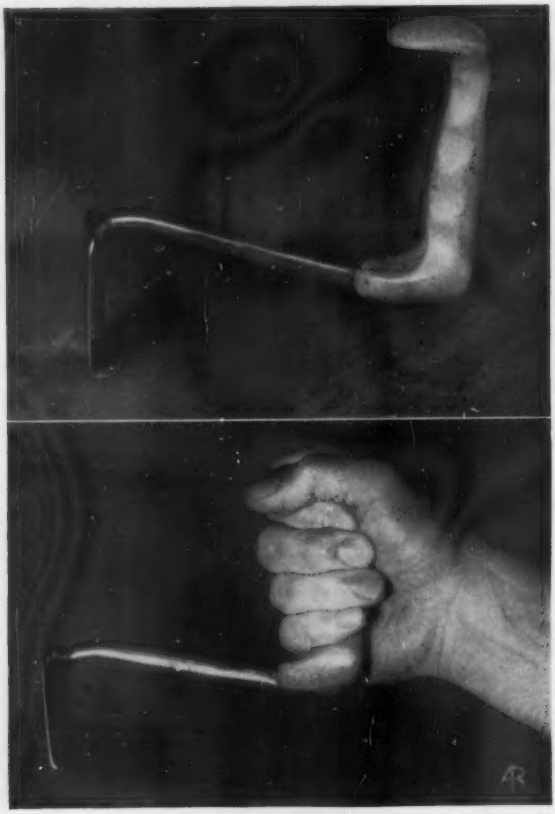
20—Visual field and cerebral dominance in relation to the act of writing. The left hand, passive and controlling, is, with the writing completed, within the left visual fields and so within the scope of the right cerebral hemisphere. The dynamic of word formation (of the ideas to come) is with the right hand and so left cerebral.

crossing of t's and the dotting of i's " in both senses!). The minor right cerebral hemisphere (and with it the left visual field of both eyes) thus appears to assume a special function in the subconscious processes of reflection, judgment and control.* Such an idea fits in with a thought given me by Sir Russell Brain in a private communication: " I have sometimes wondered whether, as you say, there are not other elements in the specialisation of the hemispheres than those with which we are familiar, and whether perhaps the left hemisphere may not be more clearly related to the ego and the right hemisphere to the external world."

In reading one must remember that semitic races, for example, work from right to left. Doubtless differentiation of cerebral hemisphere function is no more uniform for all races than it is for individuals, at least 30 per cent of whom are left handed. The writing and reading

* But we must recognise that apraxia in both left and right hands may be due to a lesion in the posterior part of the left major cerebral hemisphere and in the left hand only may be related to a lesion in the posterior communicating tracts of the corpus callosum.

of the Chinese is of special interest. Oriental characters which represent complete concepts are placed vertically. It probably is significant that when western people wish to tabulate complete concepts (for example arithmetically) they also tend to do so vertically. These matters cannot be pursued any further here, except to suggest that in the examination of pictorial material (whether aesthetic or scientific) the possibilities of a different type of cerebral differentiation of the left and right fields of vision may be of significance.



21—The pistol grip retractor. The hold is physiological and the direction of pull is efficient and comfortable.

DESIGN OF INSTRUMENTS

Enough has been said to indicate my belief that, apart from the work done, its completion has an important influence upon the craftsman. If " a bad workman complains of his tools," it is also true that the good craftsman requires the best.

Recent surgery owes much to the contribution which Sir Heneage Ogilvie has made to this subject. My own work, small as it is, was stimulated by my friend, the late H. P. Nelson, who objected to the bad design of the handles supplied by surgical instrument manufacturers: handles which no craftsman in other fields would tolerate.

As with other surgeons before me, I have adapted normal tradesmen's tools. The trouble, however, is that the addition of metallic handles often upsets the balance of such traditional

instruments and for this reason plastic fibre handles are preferable. The handles of most osteotomes and gouges, if tolerable in girth, are usually ridiculously short. It is upon the latter that the leverage of action is dependent. Like the sculptor, for precision we probably should also reduce the girth of our tools so that the hammer or mallet strikes always at the centre of the line of action. It surprises me that we " invent " new tools to perform jobs that craftsmen in other materials have done adequately for countless generations.

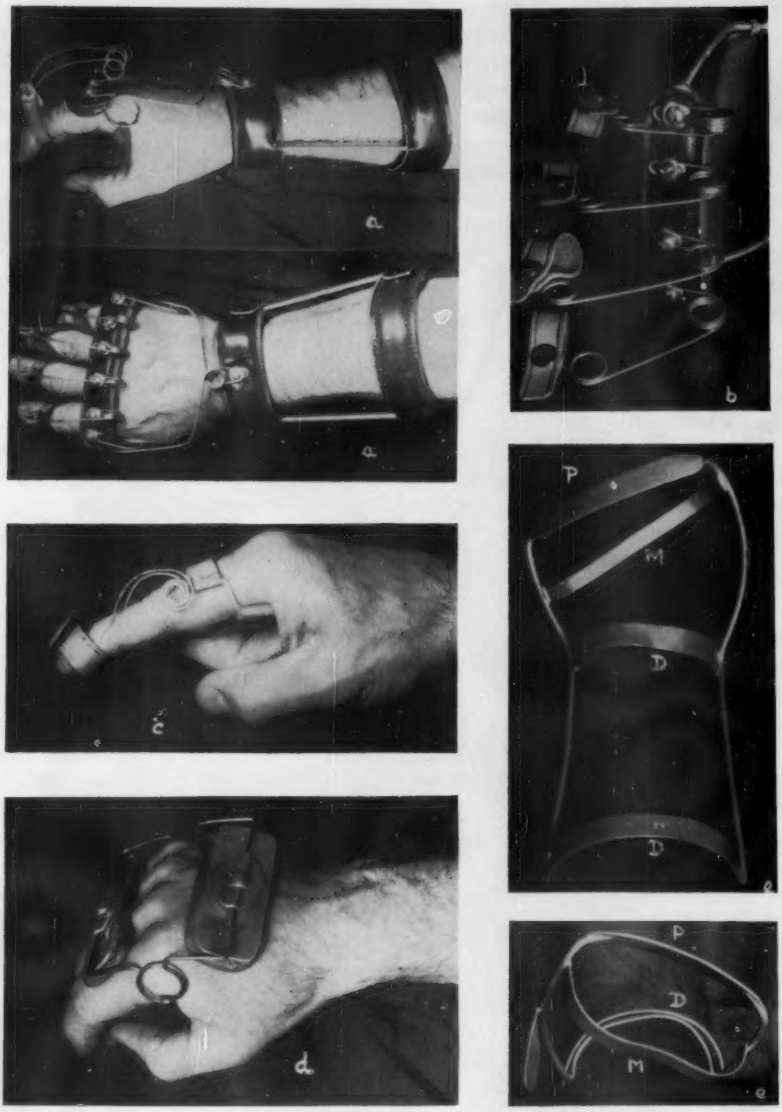
One of the chief directions in which surgeons have failed is in the design of the special instruments for their assistants, particularly retractors. Compactness and portability, which were major requirements in the equipment of the itinerant surgeon, are not so necessary today. To overcome fatigue and increase efficiency, surgeons have had to resort to self-retaining retractors based upon the design of the haemostat. Convenient as are these instruments in flat wounds with straight parallel edges, there are many occasions in orthopaedic surgery where such retractors are not efficient. Furthermore, the prolonged use of rigid metallic distraction must be damaging to soft tissues. There is reason, I think, to discourage such unsympathetic tools and to encourage intermittent distraction if this can be provided easily by a capable human assistant. Few manually held retractors permit of the economic use of the musculature of the assistant's forearm; nor do they conform to the shape of the hand. The angle of pull is rarely easy of application. I do not believe that I have yet achieved the ideal handle, but I offer one example in which the shape of the handle and the angle of hold are physiological (Fig. 21).

SPLINTAGE FOR THE HAND

In the light of what I have said, I want to add a few details of the construction of active or lively splints (Fig. 22).

Maximal sensory function is facilitated by liveliness of construction and by removing from the palmar surface all unnecessary material. Palmar support must be shaped, not only to fit its concavity, but also must have a compound obliquity which conforms to the double angle of grip (Fig. 3). Moving parts of splints should be arranged so that the axes of movement or the fulcra of levers are as far as possible in conformity, or at any rate in harmony, with the axes of limb movement. Spring action should almost always be lighter than the full power necessary to achieve a particular purpose. The physiological position of the hand should be retained by the appliance when the musculature is relaxed. For deformities of joints, as in arthritis or cicatricial contractures, lightness of spring action is equally important for the avoidance of painful stretching (and the consequent reflex inhibition of muscular action), and to safeguard the circulatory condition of distal parts. Power that is light for a normal hand is altogether too strong for the correction of deformity. Actual measurements and strengths of springs cannot be laid down. Each splint is an individual problem, but one should avoid a strong rigid corrective force of large amplitude. A small range of correction should be aimed at and the springs adjusted as this is obtained. It should be recognised that the power of the extrinsic muscles is very considerable. In my own right hand, measurements indicate that the pull of the flexor sublimis on each of the index and middle fingers can be developed to about 40 lb. On the ring and little fingers, the figures are approximately 24 and 15 lb. respectively. At the other end of the scale the power is infinitesimal. Of course, there must be great variations as between one individual and another. Nevertheless, recognition of the strong forces available will do much to explain the severity of some of the deformities seen in polyarticular arthritis.

The mechanics of spring action I do not profess completely to understand. In a somewhat rough and ready way I and my associate, Mr Fred Suter, have determined that the spring acts as a motor by torque, its helix is compressed by the weight of the limb and by the action of antagonist muscles. Relaxation of the latter causes the spring to act by the rebound from its torque. Length of spring arm provides leverage, itself modified by the elasticity of the material.

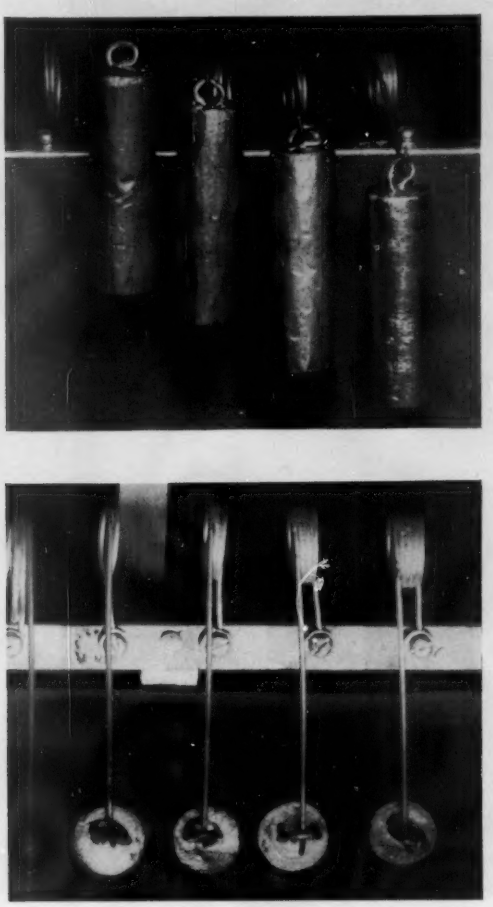


22—A variety of lively splints. *a*) The completely flexible "Suter" hand splint. *b*) Detail of method of attachment of springs to phalangeal bar and showing adaptability of lever length and multiple coiling. *c*) The "Suter" finger splint. *d*) The spring cage for intrinsic paralysis in which the framework comprises one continuous spring. *e*) Two views of basic framework before padding and before adding springs for finger extension; this frame is used where more rigid control of the wrist is necessary. P=Phalangeal controlling bar for spring attachments. M=Oblique metacarpal palmar support. D=Dorsal bands for forearm.

The strength of the spring is dependent upon the calibre of its wire, the size of helical coils and their number or length of wire (material being constant). The multiplication of coils increases elasticity and reduces power (Fig. 23).

ASSESSMENT OF INDUSTRIAL DISABILITY

Perhaps it is a sign of present day values that we find among some surgeons a tendency to look upon the hand rather as part of a machine. Kestler, for example, distinguishes four dominant functions and portrays the hand as a hook, a ring, forceps and pliers. Mervyn Evans distinguishes between the pincers and vice action of the hand. It is only fair to state



23—Experimental framework for testing spring action. Here used to demonstrate effect of multiple coils upon the power of the springs. The weights and lever lengths are exactly the same but the wire is coiled once, twice, three and four times.

that these writers are discussing the horribly mangled limbs of industry and that they would be amongst the first to recognise other values.

Possibly the craftsman is less accident prone than his mass-producing confrere. Centralised welfare schemes have to regard people as being of a common mould, if not as part of a machine. Nevertheless, the tragedy of manual disability for the craftsman is not only in the loss of earning capacity and is not to be measured in the amounts of compensation

he receives. No longer need we regard a skilled workman as any less unfortunate if he injures his *left* hand. Fortunately, it is in the realm of the intellect that real compensation is to be achieved for the craftsman, and it is along these lines that modern industrial rehabilitation has progressed so well.

OCCUPATIONAL THERAPY

Prominent in the remedial measures for the hand, and, through the hand, the whole patient, is occupational therapy. I believe the time has come to assess its value and to enquire into

24—Hands as the complement to the enquiring mind: Sir Thomas Fairbank—surgeon, teacher and perpetual student.

methods and aims. Industrial rehabilitative training I put apart, for this seems essentially good and beyond question. The reason is its integration with craft rather than with a therapeutic technique. It is thus largely related to the individual's previous experience, interest and needs. While I have great appreciation of the objects of occupational therapy there are, I think, some defects. My first objection is that it is called " therapy." The second objection is that occupational therapy is so often unrelated to the individual's experience, interest and needs. The third is that it stresses therapy as an end instead of its real purpose which should be occupation. In other words, occupational therapy should be concerned with craftsmanship, than which there is no better means or end to be found, apart from good

doctoring and nursing, in the recovery of health. Occupational therapy is educational. Training the practitioners in this field should be based upon sound principles of education and art in its widest sense and should be conducted in schools such as the Central School of Arts and Crafts in London, established under the aegis or approval of the Ministry of Education. The tradition of occupational therapy in hospitals, such as it is, is based too much upon weaving, to the disadvantage of many other fundamental crafts. Properly managed, adult occupational education in hospitals can play its part in overcoming the evils of the modern industrial age and perhaps of some of the present morbidity.

OTHER FUNCTIONS OF THE HAND

As an instrument of sensation and as an instrument of expression, there is a vast field worthy of practical and philosophic study. For example, is the strict no-touch technique altogether desirable when it involves the unnecessary manipulation of living tissues by harsh metallic tools?

The functions of the hand are indeed complex, but we do wrong if we follow those writers who regard it as a distinctly human achievement, or intrinsically as the most remarkable product of evolution. Wood Jones has stressed its essentially primitive undifferentiated nature, and Zuckermann and other writers have drawn attention to the variety of minute movements of, for example, the Rhesus monkey. Conversely, most of us will be aware of individuals born with two rudimentary lobster-like claw hands, who appear well balanced and possess considerable skill.

As an instrument of expression we should remember the relationship of the hand with speech, and the support which this gives those biologists who regard the evolution of the forelimbs into hands as a process of vicarious substitution separated from the mouth of the quadruped.

Not only in speech but in the activities of the enquiring mind, the hand is to be identified. In a paper presented in honour of a great surgical philosopher I am happy to be able to include a study of the hands and the attitude so characteristic of the great man we hold in such affection—Sir Tom Fairbank.

CONCLUSIONS

We surgeons are privileged in having a profession which also encompasses a craft; by it we should have understanding of the problems of craftsmanship in modern life. Ultimately it is at the core of human happiness. Craftsmanship is based upon creative ability, good design, the almost loving conflict of man and material, and the consequent unification of both in completion. Craftsmanship is only possible as a manifestation of individuality. Integrity and invulnerability are its outcome.

May it not be that in the atomic age (if there is one outside of destruction) there may be a return to the craftsman's life of the eighteenth century; not of necessity for the maintenance of economic life, because that will be done for him by the machine, but rather for the maintenance of health?

It remains for me to thank my colleagues, surgical and nursing, at the Princess Elizabeth Orthopaedic Hospital, Exeter, for the great assistance they have given me. I owe much also to the late Fred Suter of the Devonian Orthopaedic Workshops, a great craftsman, for his help in the mechanical problems involved, and to Mr Arthur Reaney, M.S.R., for the photography.

VOLKMANN'S CONTRACTURE: TREATMENT BY EXCISION OF THE INFARCT

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From the Institute of Orthopaedics

The purpose of this paper is to describe a method of treatment of Volkmann's contracture which is based on a conception so simple as to be self-evident. No attempt will be made to discuss the etiology of the disorder, which was established with great precision and clarity by Griffiths (1940) and is now generally recognised to be an acute ischaemia almost invariably of arterial origin. Nothing will be said about prevention. However, I must refer to the morbid anatomy and histology of muscle and nerve ischaemia because the proper application of the treatment depends on the accurate assessment, clinically and at operation, of the extent and severity of the damage.

The lesion is a massive infarct.* The treatment to be described is, briefly, excision of all irreparably damaged muscle and nerve followed by such reconstructive procedures as may be required to minimise the deficiency.

MATERIAL

Although the method is more widely applicable,† the discussion will be limited to the commonest form of ischaemic contracture, that affecting the forearm muscles. The cases described have been selected on account of their severity. It is only in such cases that excision is appropriate. I have omitted a number of cases of great severity in which the whole forearm was hopelessly damaged as a result of a proximal arterial lesion. Some were investigated in detail, but the data add nothing to what is discussed below, and reconstructive surgery had nothing to offer these unfortunate patients.

It will be observed from Table I that involvement of the main nerve trunks was frequent, in itself a sufficient indication of the gravity of the ischaemia. The first six patients were adults, all in the fighting services, who were sent to the Peripheral Nerve Injuries Centre at Oxford during the second world war. They were described in a paper by Holmes, Highet and Seddon (1944) in which evidence was presented to prove that the nerve lesions found in cases of severe ischaemia of the forearm were due to the initial ischaemia, not to the subsequent contracture of the damaged muscle. These patients were subjected to very detailed examination. The vascular lesion was investigated by oscillometry, arteriography and operative exploration. The muscles and nerves were also inspected and many pieces removed for histological examination. Although at that time we concluded that the nerve lesions were irreparable, it happens that the information obtained is now relevant to the treatment proposed. Thus there are six patients who were investigated but not treated, followed by a further ten in whom investigation was followed by more or less successful amelioration of the ischaemic damage.

* Only by this dogmatic brevity can one avoid entanglement in the labyrinthine discussions on the exact pathology of ischaemic contracture that have taken place in France. Those interested may consult Cahuzac and Jung (1946) who give a good summary of recent French work.

† We have used the method three times for ischaemia of the calf muscle. My colleague J. I. P. James has successfully excised ischaemic carpal interossei in three cases. However, the trouble in the hand is that the palmar interossei are usually more severely affected than the dorsal, and if the palmar interossei are excised the nerve supply to the dorsal muscles is destroyed.

TABLE I

THE INVOLVEMENT OF MUSCLE GROUPS AND OF NERVES IN SIXTEEN CASES OF VOLKMANN'S CONTRACTURE

Case number	Reference number	Age (*years*)	Injury	Ischaemic damage				
				Muscle		Nerve		
				Flexors	Extensors	Median	Ulnar	Radial
1	G 24	60	1940. Fracture of surgical neck of humerus, axillary wound	++	+	++	++	
2	B 21	31	1941. Fracture mid-shaft of humerus	+++	++	++	++	+
3	C 1	35	1941. Fracture mid-shaft radius and ulna	+++	++	++	++	
4	M 32	41	1941. Bomb wounds injuring lower end humerus and brachial artery	+++	++	+ also direct injury	+	
5	S 35	37	1942. Colles's fracture	++	++	++	+	+
6	G 58	26	1943. Fracture upper third radius and ulna	+	(+)	+	+	
7	16,988	9	1946. Supracondylar fracture	+++		++	++	
8	17,317 X	5	1947. Supracondylar fracture	+++	++	++	++	
9	644	5	1944. Comminuted fracture of elbow	+++		no evidence of damage six years later		
10	41,527	8	1939. Supracondylar fracture	+++		++	++	
11	46,751	7	1951. Monteggia fracture-dislocation	++		+	+	
12	55,873	6	1951. Supracondylar fracture	++		+	+	
13	62,449	29	1952. Prolonged compression of forearm	+		+	+	
14	70,344	5	1953. Transcondylar fracture	+++		++	+	
15	100,878	11	1954. Supracondylar fracture	+++	++	++	++	
16	76,895	30	1954. Comminuted fracture lower end of humerus	+++		++	+	

MORBID ANATOMY

The zone of damage differs considerably from the typical conical infarct seen in solid organs such as the lung or spleen. It differs both in shape and in the distribution of tissue change. When the distal part of the brachial artery goes into spasm as a result of severance, nipping or contusion, an ellipsoid zone of ischaemia is produced with its central point somewhere about the middle of the forearm (Figs. 1 to 3). The degree of damage is usually greatest at the centre (Table II), whereas at the periphery it may be comparatively slight, thanks to the collateral circulation. It is rare for the superficial muscles to be affected as

FIG. 1

FIG. 2

Figure 1—A schematic drawing of the zone of ischaemia in Volkmann's paralysis of a right forearm. The yellow zone is that in which complete necrosis is commonly found and it may involve the median nerve. In the pink zone the muscle may be completely or partially fibrotic. Figure 2—A lateral view of a right forearm showing how the deep extensor muscles may be involved.

much as those more deeply placed. Furthermore, a forearm muscle is often unequally affected throughout its length; its extremities may escape.

There are great variations. These depend on the level of the arterial lesion, no doubt too on the degree and duration of ischaemia and on the hazard of arterial spasm involving this or that branch of the brachial vascular tree—though neither of these factors is susceptible of investigation. There is yet another factor, though I have not been able to take account of it—the situation of the neurovascular hila of the long muscles, so elegantly described by Brash (1955).

What is so remarkable is that in spite of these variables there is indeed some sort of pattern. The ellipsoid zone of ischaemia has its axis somewhere near the anterior interosseous artery. The muscles that usually suffer most severely are the flexor digitorum profundus and the flexor pollicis longus which lie on either side of it. Moreover, if the extensor muscles are affected, the most vulnerable are those most deeply situated.

A main nerve trunk running through the centre of the ellipsoid zone of ischaemia will tend to suffer great damage. In one lying at the periphery the damage will be less severe. Thus the median nerve suffers more frequently and more severely than the ulnar nerve.

FIG. 3*a*

Figure 3*a*—The shading represents the degree of involvement of the various muscles. This diagram is based on the data given in Table II.

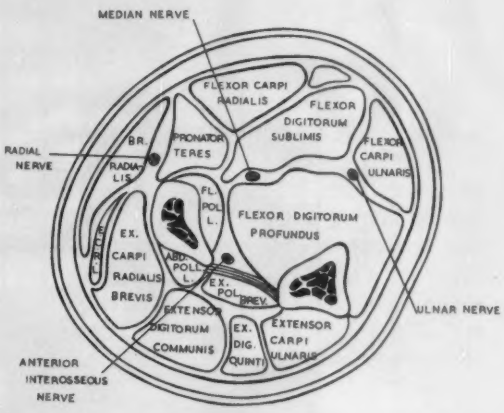

FIG. 3*b*

Figure 3*b*—Key to muscles, the plane of section being in the upper third of the forearm.

Muscle

a) *Reversible damage*—Le Gros Clark and Blomfield (1945) have shown experimentally that in the rabbit damage to muscle due to ischaemia may be reversible. Under favourable conditions the dead muscle fibres are removed and replaced by new ones which grow longitudinally along the surviving sarcolemmal tubes from the unaffected muscle above and below. In man some such process must occur more frequently than is realised. Anyone who has made even a cursory study of the histology of ischaemic contracture is familiar with the appearance of regeneration, usually abortive, at the periphery of the ischaemic zone (see Hughes 1948, Horn and Sevitt 1951, and Fig. 4). In Volkmann's ischaemia there is often at first a total paralysis of the forearm, but within two or three months the extensor muscles recover well and there may be some recovery in the flexors too. This recovery is most noticeable in the superficial muscles. Reference to spontaneous recovery will be made later in the paper.

TABLE II

THE MUSCLES AFFECTED IN SIXTEEN CASES

		CASE NUMBERS																Healthy	Partial fibrosis	Fibrosis	Necrosis	Not recorded
		1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16					
SUPERFICIAL FLEXORS	Pronator teres									NR								7		7	1	1
	Flexor carpi radialis																	6	1	6	3	
	Flexor digitorum sublimis																	3	2	6	5	
	Flexor carpi ulnaris												NR		NR	NR		5	1	4	3	3
DEEP FLEXORS	Flexor digitorum profundus																	1	1	4	10	
	Flexor pollicis longus																	1	1	5	9	
DEEP EXTENSORS	Extensor pollicis longus	NR																9	2	2	2	1
	Extensor pollicis brevis	NR	NR															9	1	2	2	2
	Abductor pollicis longus				NR													9	2	2	2	1
	Extensor indicis			NR		NR												9		2	3	2
SUPERFICIAL EXTENSORS	Brachio-radialis																	15	1			
	Extensor carpi radialis longior																	15	1			
	Extensor carpi radialis brevior		NR	NR														13	1			2
	Extensor digitorum communis																	13		2	1	
	Extensor carpi ulnaris	NR	NR	NR														12		1		3

= Healthy
= Partial fibrosis
= Fibrosis
= Necrosis
NR = Not recorded

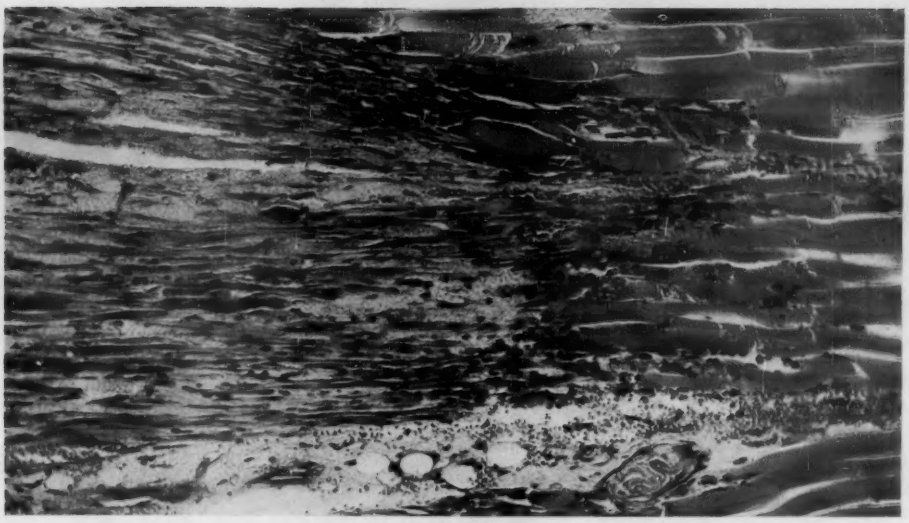

FIG. 4
The tissue adjacent to the necrotic muscle contains slender regenerating muscle fibres. (×145.)

b) *Partially reversible damage*—Some muscles show partial recovery but develop a contracture. They exhibit a certain pallor and, histologically, a diffuse fibrosis.
c) *Irreversible damage*—There is every gradation of fibrosis up to almost complete fibrous replacement of a muscle with little contractile tissue surviving (Figs. 5 and 6). The clinical manifestation is contracture and an absence of contractility. Lastly, a zone of muscle may undergo complete necrosis which leads to the most dramatic changes. The muscle is converted into a greenish-yellow pulpy mass, completely bloodless, which is easily scraped out with a spoon. Histological examination of this material confirms that it is necrotic (Fig. 7). Yet here again there is a gradation. There may be minute foci of necrosis in an otherwise completely fibrotic muscle (Fig. 8).

In a typical severe case the centre of the infarct is dead tissue. The zone of necrosis is surrounded by one in which necrosis and fibrosis occur together. Much of the necrotic material is removed by phagocytosis (Figs. 9 and 10). More peripherally the fibrosis is less pronounced and in the outermost zone there is reversible ischaemia.

Nerve

There are similar changes in the nerves which have been fully described by Holmes *et al.* (1944).
a) *Reversible damage*—Wallerian degeneration can occur as a result of mild ischaemia and it may be followed by the usual type of regeneration of fibres from the proximal healthy part of the nerve.
b) *Irreversible damage*—A nerve may undergo complete collagenisation in every respect comparable with total fibrosis of a muscle, and this process is irreversible. It is easily recognised histologically (Fig. 11) and is manifest at operation as a gross shrinkage of the nerve trunk, which may be less than a quarter of its normal diameter (Figs. 6, 12 and 13).
c) In the centre of a zone of total ischaemia a nerve may share in the massive necrosis (Fig. 7) and be converted into a dull yellow avascular structure which retains its axons and myelin sheaths simply because there is nothing to remove them.

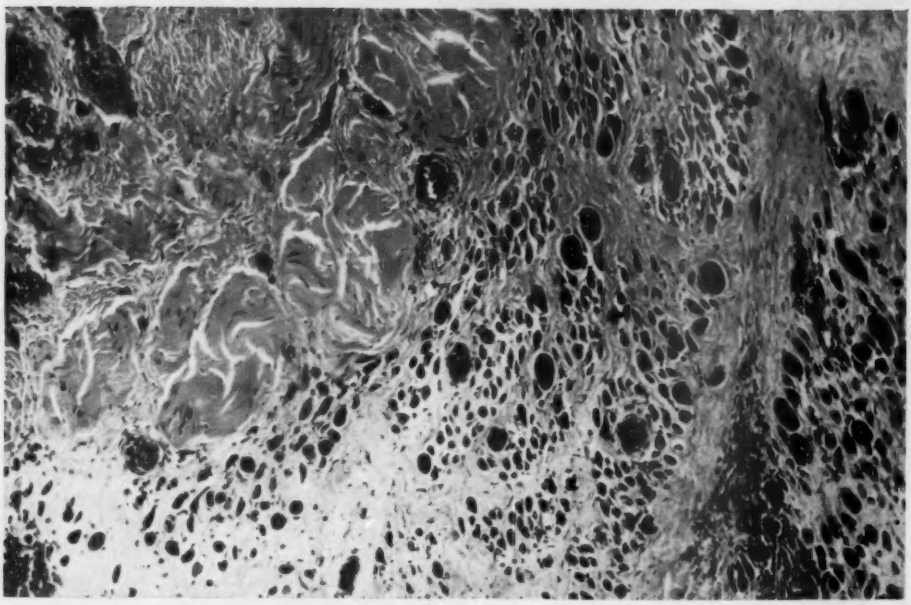

FIG. 5
Atrophy and fibrosis of non-necrotic muscle. (×145.)

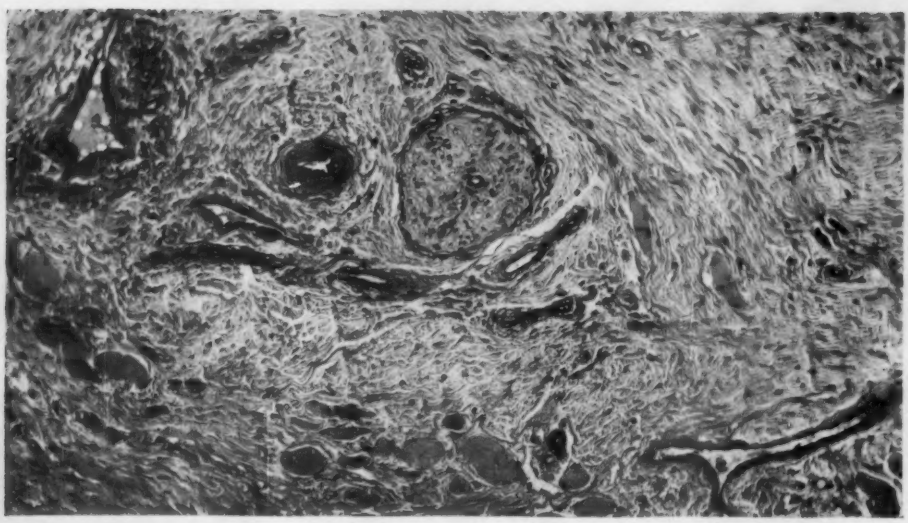

FIG. 6
An area of intensely fibrotic muscle. The muscle fibres included in the field are not necrotic. A small nerve bundle shows severe loss of myelinated fibres with some endoneurial fibrosis. (×145.)

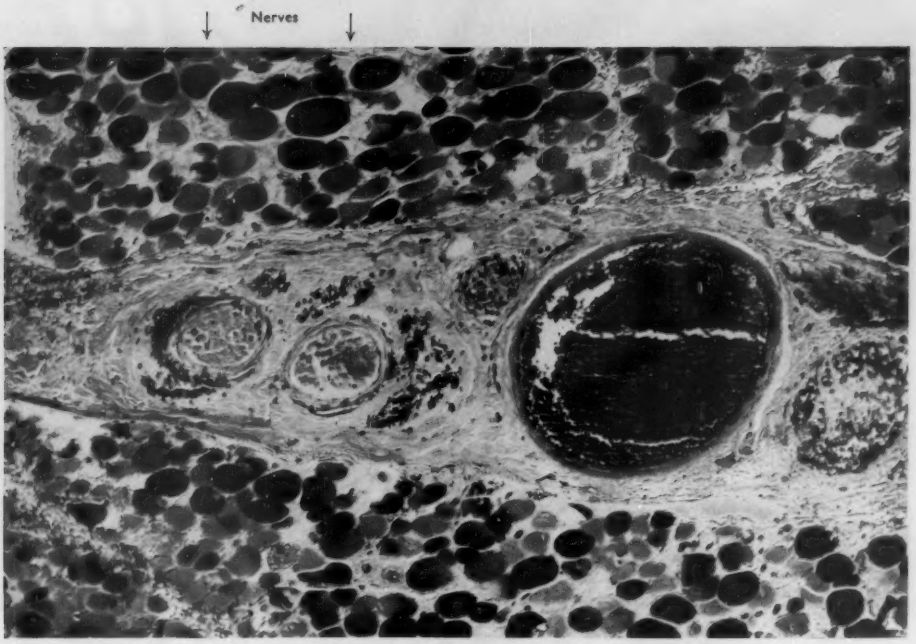

FIG. 7

The centre of a necrotic zone. Muscle fibres are all necrotic; note their variable staining reaction. The artery in the centre of the field is distended with blood clot and its wall is necrotic. Two nerve bundles are completely necrotic. The only persisting cells in this field are a few fibroblasts in the connective tissue surrounding the vessels and nerves. (×145.)

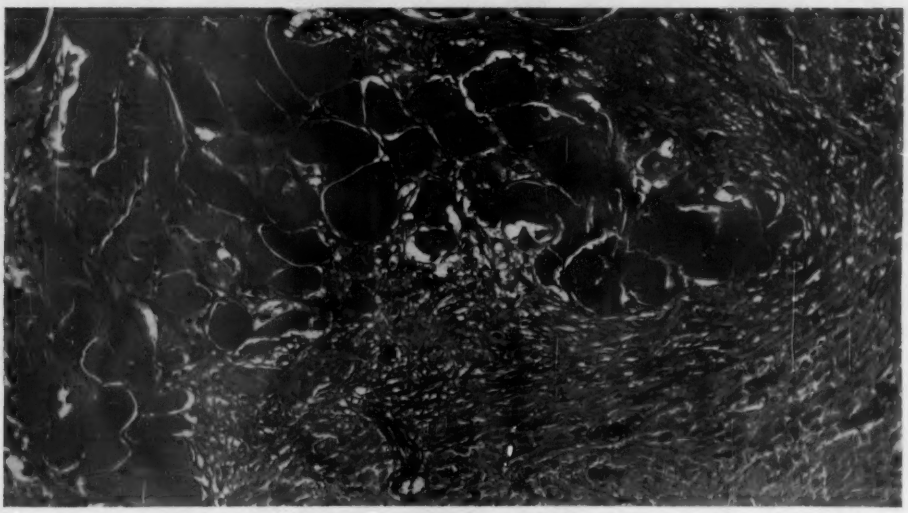

FIG. 8

An island of necrotic muscle fibres surrounded by newly formed tissue. (×145.)

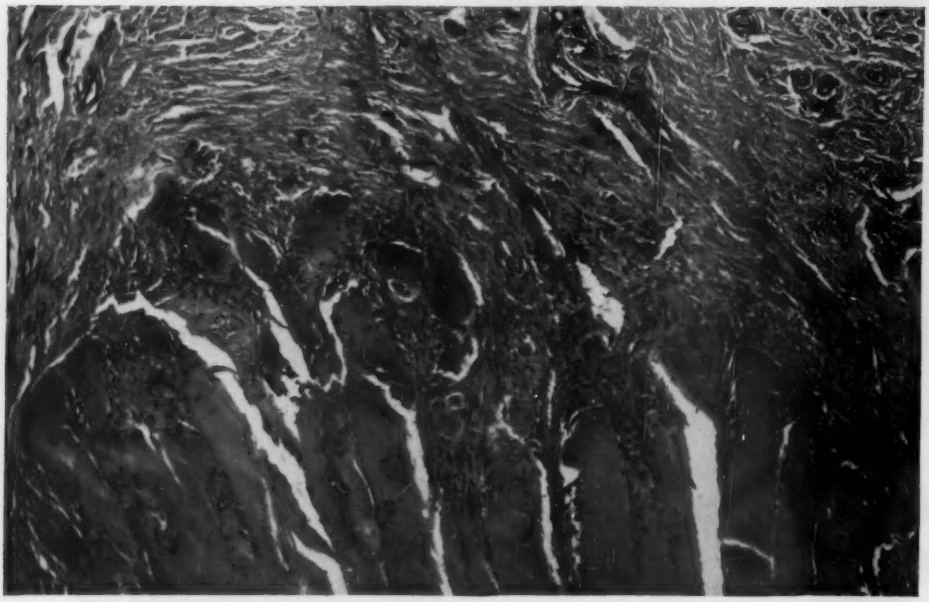

FIG. 9
Phagocytosis of the necrotic fibres by macrophages and by multinucleated giant cells. (×145.)

FIG. 10
Higher power view of area of phagocytosis. Note the accentuated cross-striation in the necrotic muscle. (×255.)

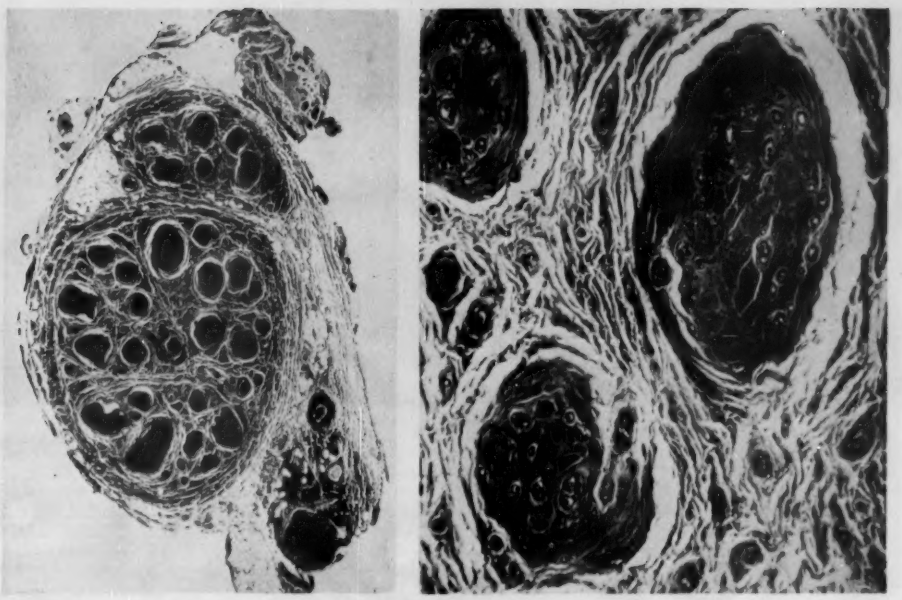

FIG. 11

Nerve bundles completely destroyed by interstitial collagenisation. *Left*, low magnification. *Right*, high magnification. The Schwann tubes are almost completely replaced by dense collagen. Magnification not recorded.

SPONTANEOUS RECOVERY AND CONSERVATIVE TREATMENT

Excision of the infarct is an operation to be employed only in severe cases of Volkmann's ischaemia. Although the structural changes that are reversible are easily distinguishable pathologically from those that are not, the clinical distinction is difficult and one can make it only by waiting and watching.

Here is the case history of a boy on whom I twice decided to operate because of the great severity of the paralysis. On each occasion operation was postponed because just when the situation seemed very gloomy, he exhibited a little return of function. In the end recovery was excellent with only a minimal contracture.

C. C., a boy aged five years, suffered a lesion of the right brachial artery from a supracondylar fracture. Vigorous efforts to restore the circulation were unsuccessful. He developed paralysis of all muscles below the elbow and sensibility was lost in the median, ulnar and superficial radial areas. Recovery occurred as follows:

a) in the extensor group (Table III) it began on the fiftieth day and there was no "march" of recovery from the proximal to the distal muscles; *b*) in the flexor group recovery began on the eighty-seventh day and here again there was no regular proximal to distal progression; *c*) the intrinsic muscles of the hand, which were paralysed as a result of the nerve lesions and showed a reaction of denervation, began to recover much later, towards the 200th day; *d*) in the palm of the hand sensibility began to return on the 140th day.

Thus there were two phases of recovery; in the forearm muscles fairly early recovery, earlier in the extensors than in the flexors, and occurring at about the same rate in all muscles; and in the hand a late recovery, resembling that following an ordinary degenerative nerve lesion in that the rate was roughly one millimetre a day for sensory fibres and rather less for motor. The sensory recovery clearly exhibited a proximal to distal progression. In another similar case recovery began at three months and was well advanced by the fifth.

TABLE III

THE PROGRESS OF SPONTANEOUS RECOVERY IN CASE C. C.

Muscles in order of innervation	Days after injury									
	46	50	60	65	87	114	153	198	212	336
Brachioradialis				2	3	4	4	4	4	4
Extensor carpi radialis longior		1	2	2	3	3	3	3	3	4
Extensor carpi radialis brevior		1	2	2	3	3	3	3	3	4
Extensor digitorum communis		1	2	3	3	4	4	4	4	4
Extensor carpi ulnaris								3	3	4
Abductor pollicis longus			2	3	3	4	4	4	4	4
Extensor pollicis longus			2	3	3	4	4	4	4	4
Extensor pollicis brevis						1	3	4	4	4
Pronator teres						2	2	2	3	4
Flexor carpi radialis					1	1	2	2	3	4
Flexor digitorum sublimis					2	2	3	3	3	4
Flexor pollicis longus					2	2	2	2	3	4
Flexor digitorum profundus (lateral)							2	2	3	3
Flexor carpi ulnaris					2	2	2	2	3	4
Flexor digitorum profundus (medial)							2	2	3	4
Thenar muscles								3	4	4
Hypothenar muscles								1	2	3
Interossei										3

1—Flicker.
2—Movement only with gravity eliminated.
3—Movement against gravity but not against resistance.
4—Movement against gravity and resistance.

It is impossible to distinguish recovery of ischaemic muscle due to regeneration of muscle fibres from that due to regeneration of the nerve fibres they contain, but the pattern of the proximal recovery in the patient C. C., and perhaps its rate, suggests that it is the regeneration of the ischaemic muscle that sets the pace. Peripheral nervous tissue is manifestly less susceptible to ischaemia than muscle, and it is conceivable that in the relatively mild ischaemia suffered by C. C. the small motor branches to the proximal muscles never underwent Wallerian degeneration, although the more peripheral parts of the large nerve trunks did so.

The important practical lesson to be learned is that one should *wait for at least three months* for evidence of spontaneous recovery in the forearm muscles before embarking on the drastic treatment proposed in this paper. During this time treatment should be limited to:

(*a*) splinting to minimise the contracture. The splint I have employed (Fig. 14), a refinement of one devised by Michel (1924), is very useful for this purpose; (*b*) movements of all joints: if mobility is lost the prognosis is very poor; (*c*) if there is a nerve lesion, daily galvanic stimulation of the muscles of the hand (Jackson 1945, Jackson and Seddon 1945).

FIG. 12

Case 7—The median nerve. The proximal part is to the right and it will be seen that it gradually tapers to a narrow strand about a quarter the normal size. Then, just distal to the small swab, the nerve enlarges again though not to the same size as proximally. The narrow part is irreparably damaged. The broader distal part showed ordinary Wallerian degeneration. A nerve-pedicle graft was carried out in this case with an excellent result.

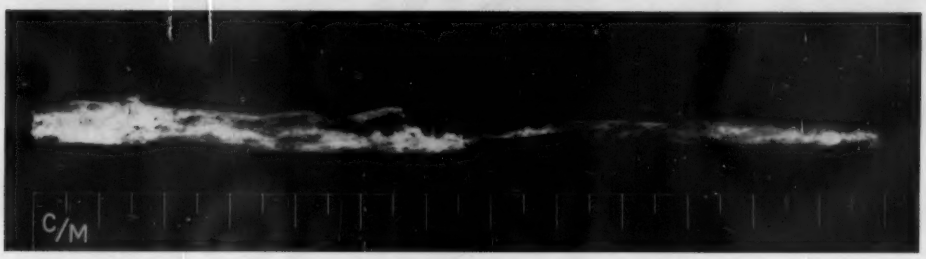

FIG. 13

Case 15—The excised segment of ischaemic nerve.

THE EVOLUTION OF THE OPERATION

The operations commonly employed for the relief of Volkmann's contracture are summarised in Table IV. They all have shortcomings and there is no recognised remedy for severe ischaemic nerve lesions.

In meditating on this problem I recalled a young man who in November 1942 lost the greater part of the tissues on the front of the forearm in a machine accident. After the usual treatment of the primary wound the raw area was covered with skin. A free graft from the ulnar nerve was used to bridge the large gap in the median; the wrist was fixed by arthrodesis and the extensors used to activate the digits (for details see Seddon 1947, Case S.63, page 161). This man made a good recovery and returned to work. He was far better off than the average patient with severe Volkmann's contracture. Why not then attempt to convert the arm damaged by ischaemia into something like one that had suffered a traumatic loss of substance? The idea seems simple enough but it took shape very slowly and the earlier operations were not very well conceived (see Tables VA and B). At that time I was unaware that Massart (1935) had corrected a contracture by excision of the ischaemic muscle.

Case 7 was the first. The median nerve (Fig. 12) was successfully repaired by an ulnar nerve-pedicle graft (Strange 1947). But the contracture was dealt with only by division of tendons at the wrist, all the muscles being paralysed. I lacked either the wit or the courage

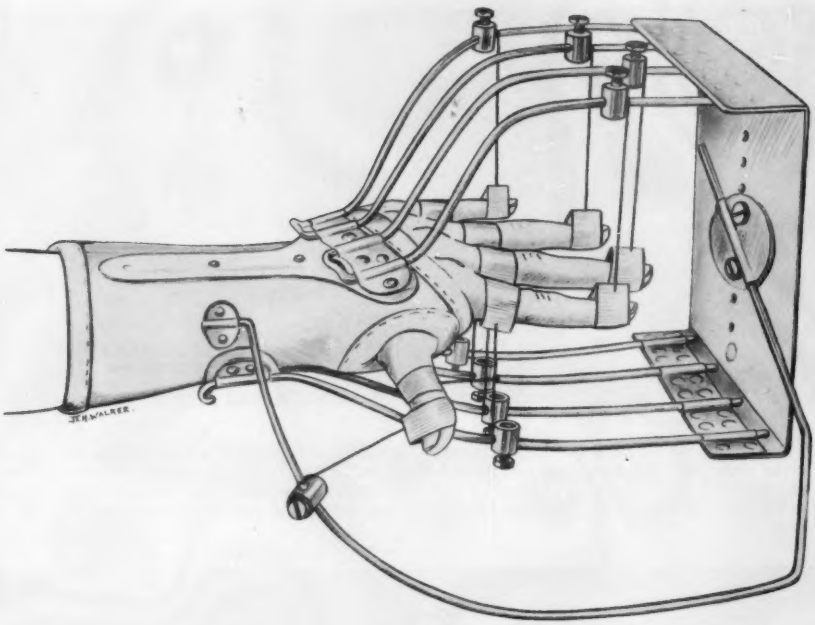

FIG. 14

The author's splint for the correction of moderate degrees of ischaemic contracture. The apparatus is cumbersome but has the merit of being removable; there is no risk of stiffness developing in any of the digital joints.

TABLE IV

A SUMMARY OF THE MAIN FEATURES OF OPERATIONS COMMONLY USED IN THE TREATMENT OF SEVERE ISCHAEMIC CONTRACTURE

Operation	Advantages	Disadvantages
Muscle slide	No interference with tendons at level of the wrist	Ineffective for paralysed muscles; all flexor muscles are treated alike irrespective of degree of damage.
Excision of proximal row of carpal bones	A minor operation; leaves soft parts relatively undisturbed	As for muscle slide; only limited correction possible
Carpectomy—Griffiths (1940), White and Stubbins (1944)	A minor operation; leaves soft parts relatively undisturbed	As for muscle slide; mutilating
Bone shortening	Leaves soft parts relatively undisturbed	Risk of non-union; ineffective for paralysed muscles; all flexor muscles treated alike irrespective of degree of damage; extensors unduly slackened; further shortening of an already shortened forearm
Arthrodesis of wrist	Leaves soft parts relatively undisturbed	As for bone shortening, except that there is no risk of non-union
Tendon lengthening	Can be applied selectively	Ineffective for paralysed muscles; risk of tendons adhering to each other
Tenotomy of destroyed muscles and tendon transplantation (Parkes 1951)	A rational operation and the only one in this list which aims at restoring power	Risk of recurrence of the contracture because the fibrotic and necrotic muscles have only been tenotomised

TABLE Va

OPERATIONS ON MUSCLE

Number	Excision of muscles	Interval after injury	Muscles divided	Interval after injury	Tendon transplantation	Interval after injury	Supplementary operations	Comments and results
7			All flexors	11 months	Wrist extensors into F.D.S.	12 months	Arthrodesis of wrist	Poor: much glueing together of flexor tendons after division from their muscles
8	All flexors	19 months	All flexors	6 months	E.C.R.L.→F.D.S.	24 months	Arthrodesis of wrist	Fair: much glueing together of tendons of F.D.P. after division from muscle
9	F.C.R., P.L., F.D.S., F.D.P.	6 years			F.C.U.→F.D.P.	74 months	Excision of proximal row of carpal bones	F.P.L. was adherent, but tenodesis effect was good. Tendon transplantation a little too tight, hence bone operation. Final result excellent
10	F.D.P.	12 years	All other flexors	12 years	Offered but never performed		Excision of proximal row of carpal bones	All procedures carried out at one session. Good correction of deformity
11	F.D.P., F.P.L.	3 months			F.D.S.→F.D.P.	17 months		Excellent
12	F.C.R., P.L., F.D.S.	3 months						Excellent
13	F.D.P.	5 months	F.P.L. lengthened	5 months				Perfect but for loss of power in F.D.P.
14	F.C.R., F.D.S., F.D.P., F.P.L.	6 months			E.C.U.→F.D.P. E.C.R.L.→F.P.L.	19 months		Excellent
15	F.D.P., F.P.L., F.C.R., F.D.S., F.C.U.	1½ months 3 months			E.C.U.→F.D.P. E.C.R.L.→F.P.L.	11 months		Too early to assess
16	F.D.S.	9 months	F.P.L.	9 months	F.C.R.→F.P.L. F.C.U.→F.D.P.	9 months		All procedures carried out at one session. Good result

F.C.R.—Flexor carpi radialis.
P.L.—Palmaris longus.
F.D.S.—Flexor digitorum sublimis.
F.D.P.—Flexor digitorum profundus.
F.P.L.—Flexor pollicis longus.
F.C.U.—Flexor carpi ulnaris.
E.C.U.—Extensor carpi ulnaris.
E.C.R.L.—Extensor carpi radialis longus.

TABLE Vb

REPAIR OF NERVES

Case number	Operation	Interval after injury (*months*)	Comments	Results
7	Nerve pedicle: ulnar to median	8 and 10		Thenar muscles very strong: pain and light touch appreciated throughout median area
8	Nerve pedicle: ulnar to median	8 and 12		Thenar muscles very strong: pain and light touch appreciated throughout median area
14	Free graft: main motor division of median into median in forearm	6	Repair carried out at same time as excision of muscle: mobilisation required for approximation	Thenar muscles strong except for abductor which is still paralysed: pain and coarse touch appreciated throughout median area. Still time for further recovery
15	Nerve pedicle: ulnar to median	1½ and 3	Repair carried out at same time as excision of muscle	Recovering steadily but final assessment not yet possible
16	Free graft: anterior interosseous and medial cutaneous of forearm into median in forearm	12	Mobilisation required for approximation	Recovering steadily but final assessment not yet possible

to excise the ischaemic mass, and although a transplantation of the wrist extensors into the flexor tendons of the digits was performed, the result was disappointing. The tendons at the wrist became glued together, a consequence of the progressive foreign-body reaction round the ischaemic zone in the forearm.

Much the same procedure was followed in Case 8, though here I attempted to make amends by excising the ischaemic mass after simple division of the flexor tendons had failed. It was a little too late. The fibrotic process glued the tendons together at the wrist and the range of movement resulting from the subsequent tendon transplantation—though the power was good—was only one-fifth of the normal (see Fig. 15).

Case 9 was easier. There was no nerve lesion. Excision of the ischaemic muscle and tendon transplantation sufficed. Even at six years after the injury there was little fibrosis round the tendons at the wrist, perhaps because the ischaemia was more proximal than usual.

In Case 10 no operation was performed until twelve years after the injury. A calcified necrotic flexor digitorum profundus was excised, the tendons of the other fibrotic muscles being divided. Because there was a fixed bony deformity at the wrist the proximal row of the carpal bones was excised. There was severe nerve damage but it was too late to contemplate any sort of repair. The patient was so satisfied with the correction of the deformity that he has not yet agreed to submit to the tendon transplantation that should give him a still better hand.

In Case 11 (Fig. 16), treated by my colleague J. I. P. James, recovery was excellent; the operation was performed early and there was no serious nerve lesion. Case 12 (Fig. 17), treated by A. F. Rushforth, was unusual in that flexor sublimis digitorum suffered gross ischaemia whereas the deep flexors escaped. Apart from loss of the superficial flexor this child now has a normal hand. In Case 13 (Fig. 18) excision of the ischaemic deep flexors was performed early. The result was perfect apart from loss of power in the long flexor of the thumb and the deep flexors of the fingers.

FIG. 15*a*

FIG. 15*b*

FIG. 15*c*

The result in Case 8. *a*—The hand before surgical reconstruction. *b*—After a nerve-pedicle graft the patient recovered good sensibility in the median area and strong power in the short muscles of the thumb which are shown opposing the thumb to the index finger. *c*—The hand is shown open. The range of movement was far short of normal but it was powerful and the patient was content.

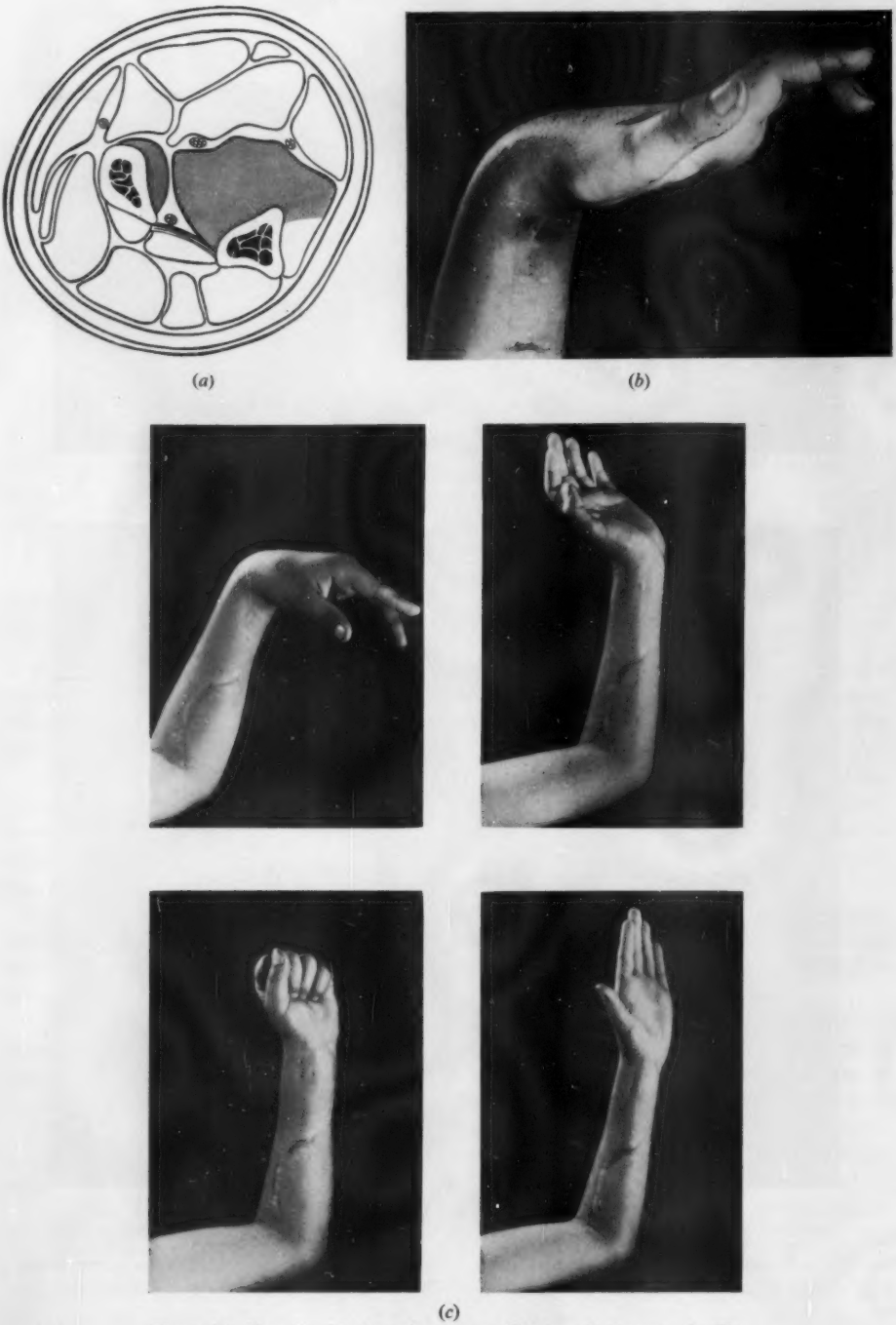

Figure 16. Case 11—Mr James's case. *a*—The muscle damage. *b*—The condition before operation. *c*—After operation. Function was perfect except for loss of power in the long flexor of the thumb.

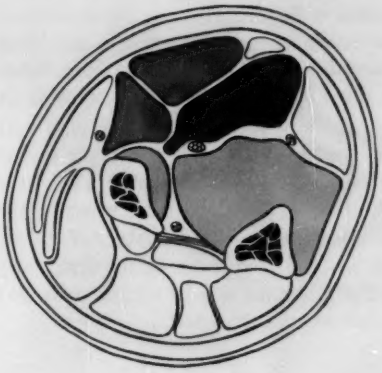

FIG. 17*a*

FIG. 17*b*

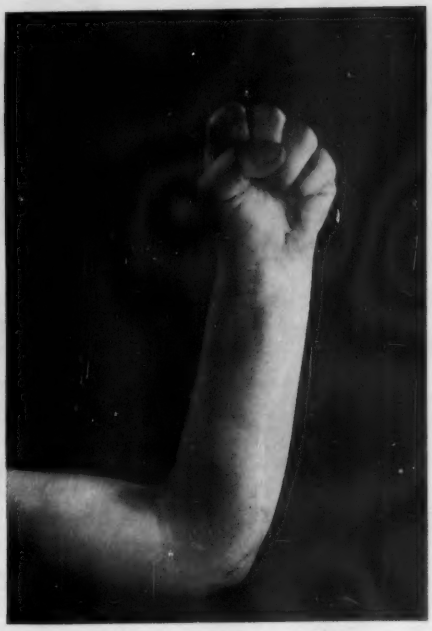

FIG. 17*c*

Case 12—Mr Rushforth's case. *a*—Muscle damage. *b and c*—Condition after operation. Function was perfect.

Of the remainder only Case 15 requires special mention. This patient, whose treatment is not yet concluded, came under my care five weeks after his injury. By this time the contracture was pronounced, there was a complete ischaemic lesion of both ulnar and median nerves, complete paralysis of all the long flexor muscles (Fig. 19) and considerable weakness of the extensors. I had been so impressed with the evil consequences of delay that I decided to explore the forearm six weeks after the injury, confident that I could recognise which structures were irreparably damaged. They were to be removed, the deformity would be immediately corrected, progressive fibrosis would be avoided and the reconstruction of muscles and nerves could begin forthwith. At operation the deep flexor muscles were found to be almost

completely necrotic in the middle of their length (Fig. 20) and were excised. But I was uncertain about the superficial flexors—they were very shrunken, pale pink and shortened, and they contracted feebly only on powerful direct electrical stimulation. Were they irreparably damaged? It was too early to be certain. Fortunately there was no need to be disturbed about them because the ischaemia of the median nerve was extreme and extended well above the common flexor mass. There was clearly no hope for these muscles and the first stage of an ulnar to median nerve-pedicle graft was performed; but it might have been most awkward. Histological examination of these doubtful muscles did, in fact, reveal grave and irreversible changes. Yet, as it was, it was necessary to wait a year for recovery of the extensor muscles before using some of them for transplantation into the flexors of the digits.

It is therefore wrong to carry out this radical treatment within three months from the time of injury and it is probably wiser to wait six months.

FIG. 18
Case 13—The circumscribed muscle damage.

THE TECHNIQUE OF THE OPERATION

The exposure and excision—The primary operation begins as a very extensive biopsy. The incision should be a longitudinal one in the mid-line of the forearm starting at the bend of the elbow and ending just above the wrist. Only in exceptional cases, such as Case 13 (Fig. 18) where the zone of ischaemia was very circumscribed, should a shorter incision be used. A bloodless field is essential during the first forty-five minutes but the pressure bag should then be released. It is useful to expose the median nerve at the bend of the elbow so that it can be stimulated directly and the contractility of the forearm muscles observed at intervals throughout the operation.

The superficial muscles are examined first. If they are necrotic they should be excised. If they are pale and fibrotic, and if they contract extremely feebly or not at all when the nerve is stimulated, they should be excised. In either case the dissection begins distally after identification of the median nerve, which must be traced proximally with the greatest care. However, certain muscles although shortened may contract well, in which case a decision must be made as to whether their tendons should be lengthened near the wrist or, if they are not flexors of the fingers, used for transplantation if all the finger flexors prove irreparably damaged. So they are best left alone until the state of the deep muscles has been determined.

The deep muscles are brought into view immediately when the superficial ones are excised. If the superficial muscles are to be preserved, then the deep ones are best exposed by an

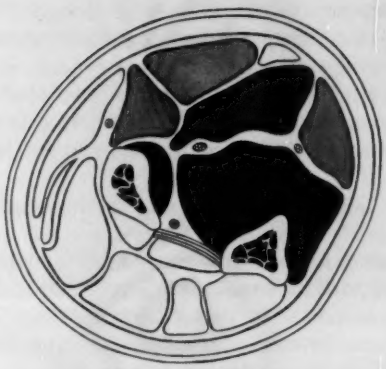

FIG. 19
Case 15—The muscle damage.

FIG. 20
Case 15—The excised mass of necrotic muscle, flexor digitorum profundus and flexor pollicis longus.

incision which again starts distally between the tendons of flexor carpi radialis and flexor digitorum sublimis. This exposure enables the surgeon to follow the median nerve proximally. More often than not the deep flexors of the fingers and the flexor pollicis longus are largely necrotic and should be excised *en bloc* from the point where the tendons make their appearance up to the junction—if there is one—of necrotic with healthy muscle. When the excision of these muscles has been completed, most if not all of the contracture of the digits will have been straightened out. The remainder of the contracture will disappear when the superficial muscles have been released. When both the superficial and the deep flexors of the fingers are hopelessly damaged, the tendons of the superficial flexors should be removed from the carpal tunnel; likewise if the superficial flexor alone is excised.

The surgeon now turns his attention to the nerves. If the ischaemia has been very severe it will be found that the median nerve narrows gradually from a point near the upper limit of the ischaemic zone until its diameter is about a quarter or a third of the normal. Its colour is greyish-yellow and if the tourniquet is released it will be found that this reach of the nerve is avascular. At the lower limit of the ischaemic zone the nerve gradually swells out again,

but not to its normal diameter. Below the zone of ischaemia it has undergone ordinary Wallerian degeneration and has the appropriate diameter of about two-thirds of the normal.

The ulnar nerve is then inspected if there are signs of total interruption, but the degree of ischaemia is never as great as in the median. However, unless the diameter of the ulnar nerve and its colour and consistence are normal, it may safely be assumed that this nerve likewise has suffered irreparable damage. If the nerve appears and feels almost normal, it should of course be left alone.

Reconstructive procedures. *a) Muscle*—If the deep flexors alone require excision, surgical reconstruction is not essential. However in James's case (11) he aimed at and achieved a superb result by transplanting flexor digitorum sublimis into the tendinous remains of the deep flexor. The loss of flexion of the terminal phalanx of the thumb is relatively unimportant. If all the flexors of the digits have to be excised, then the surgeon must borrow what he can for transplantation into them. There may be a fairly powerful flexor carpi ulnaris (Case 16). Flexor carpi radialis is unlikely to be of any use where flexor digitorum sublimis has had to be removed. He may need to borrow one or more of the wrist extensors and here another problem arises. In a case of such severity it is possible, indeed likely, that the extensor muscles are feebler than normal. It is then wiser (Case 15) to wait a few months for the extensors to regain their strength before using them to activate the digits.

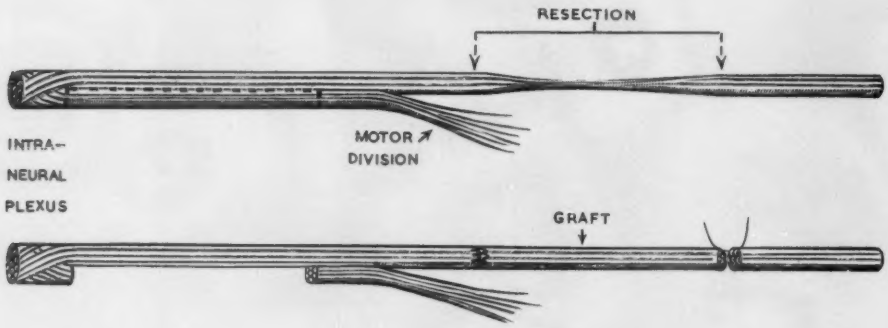

FIG. 21

A method of free grafting that can be used for repair of the median nerve in Volkmann's ischaemia. The intraneural plexus, above which dissection must not extend, is not visible on the surface as shown here but is easily apparent when the graft is being prepared. The graft should be about 15 per cent longer than the gap to be closed in order to allow for shrinkage. If it fits exactly the graft should be relaxed by flexion of the elbow.

b) The median nerve—If both the ulnar and median nerves have suffered severe ischaemia, a nerve-pedicle graft (Strange 1947) should be employed. There will of course be a permanent ulnar paralysis, but the quality of the recovery in the median has been so good in my three cases that this radical operation is fully justified.

Where the ulnar nerve appears healthy, the only possible solution is a free graft and the difficulty then is to find enough material. However, in such a case the gap to be bridged will not be very extensive, and on the two occasions (Cases 14 and 16) when free grafts were used sufficient material was found. There are two local sources of nerve grafts. The medial cutaneous nerve of the forearm will yield one and perhaps two strands if resected from the bend of the elbow up to the axillary fold. Although these grafts are long enough, their calibre is insufficient. One may then be compelled to employ also the motor division of the median itself when, as is likely, all the forearm flexors have been irreparably damaged (Fig. 21). The preparation of this graft calls for a delicate and tedious dissection.

It will be seen from Table V that the nerve and tendon reconstructions have usually been carried out at different times, though one cannot generalise about which should come first.

Lastly, in a severe case, it may be necessary to perform an arthrodesis of the wrist if destruction of the flexors and borrowing extensors for activation of the flexors of the digits has caused loss of control of the wrist.

RESULTS

The results of repair of ischaemic nerves require little comment. The procedures used, although delicate, are technically straightforward and no mistakes were made in the five grafting operations that were performed. In three recovery has been exceptionally good; sensibility throughout the median area returned to the same extent as is seen after a satisfactory suture of the median nerve at the wrist, and the thenar muscles regained power little short of normal. In the other two cases final assessment is still not possible but recovery appears to be proceeding satisfactorily. Of the remaining five cases in which no repair was performed, there was one (Case 10) in which the lesion was of such long standing that nothing could be done. In another (Case 9) there was no evidence of nerve damage and in the other three recovery occurred spontaneously.

As has already been indicated the operations performed on muscles gave indifferent results in the earlier cases; this was before a more radical policy had been evolved. Where the tendons of hopelessly damaged muscles were simply divided there was a tendency for the deformity to recur, but in none has it done so to an extent that was crippling. All the patients were satisfied with their hands *qua* deformity. Where excision was performed early, as in Cases 11 to 16, correction of the deformity was very good and in four out of six cases complete. When excision was performed late, as in Cases 9 and 10, excision of the proximal row of the carpal bones was required as a supplementary operation.

The tendon transplantations need not be considered in detail. It is sufficient to say that in the seven cases in which they were performed the results were satisfactory except when the ischaemic muscles had been dealt with by simple tenotomy.

SUMMARY

1. In the common type of Volkmann's ischaemic contracture affecting the forearm flexors, the infarct takes the form of an ellipsoid with its axis in the line of the anterior interosseous artery and with its central point a little above the middle of the forearm. The greatest damage is at the centre and usually falls most heavily on flexor digitorum profundus and flexor pollicis longus, which are often necrotic. Those muscles more superficially placed, and sometimes the deep extensors, are more likely to exhibit fibrosis.

2. The median nerve runs near the centre of the ellipsoid and may exhibit profound ischaemia. The ulnar nerve, lying at the edge of the ischaemic zone, tends to be less severely affected.

3. The treatment for this condition is excision of all tissues irreparably damaged by ischaemia. If this operation is performed within twelve months from the time of injury, correction of the contracture should be almost complete. The tendons of shortened but active muscles are lengthened or transplanted.

4. After such excision it is possible to carry out reconstructive procedures commonly used in the surgery of lower motor neurone disorders and of trauma. A wide variety of tendon transplantations is available. The median nerve may be repaired either by a free graft or, in cases where both nerves have been extensively damaged by ischaemia, by an ulnar to median nerve-pedicle graft.

Mr J. I. P. James and Mr A. F. Rushforth have generously allowed me to include Cases 11 and 12 respectively in this series. The histological work in all the recent cases has been carried out by Dr H. A. Sissons, Morbid Anatomist at the Institute of Orthopaedics, and I am grateful to him for his lively interest in this aspect of the work and for the excellent photomicrographs. Mr D. M. Brooks and I have carried out the various operations that were performed and he has contributed considerably to the evolution of the policy here recommended. Mr D. Lloyd Griffiths has been kind enough to read and criticise the manuscript.

Figures 3*a* and *b*, 16*a*, 17*a*, 18 and 19 are based on Figure 315 in *Angewandte und topographische Anatomie* by G. Töndury, 1949, Georg Thieme Verlag, Stuttgart, for the use of which I am indebted to the author and publishers.

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TENDON GRAFTS FOR FLEXOR TENDON INJURIES IN THE FINGERS AND THUMB

A Study of Technique and Results

R. GUY PULVERTAFT, DERBY, ENGLAND

The object of this work is to report the results obtained in a consecutive series of 149 cases of flexor tendon grafting in the fingers and thumb, and to study the lessons which failure and success have taught. The first graft was performed in 1942 and the period under review includes the year 1954. No case has been omitted except where an adequate late assessment has been impossible, or for some special reason which will be stated. Three groups of tendon divisions are considered: 1) flexor digitorum profundus beyond the distal palmar crease; 2) flexor digitorum profundus and sublimis in the same region; and 3) flexor pollicis longus beyond the wrist joint.

It is unsatisfactory to classify results as good, fair and poor because these terms convey little meaning. A method is needed which is accurate without containing excessive detail and which can be readily understood. In 1950 Boyes published a critical evaluation of the

FIG. 1

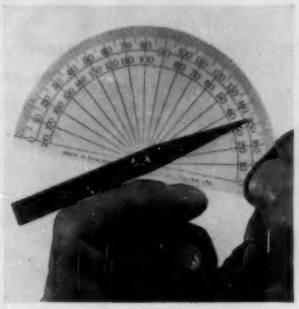


FIG. 2

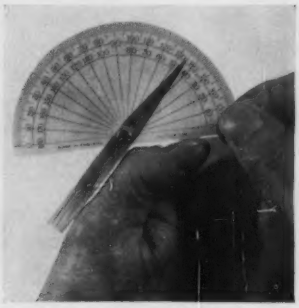


FIG. 3

Figure 1—Measurement of finger flexion. The distance from the pulp of the finger tip to the distal crease of the palm is here one inch. Figure 2—Measurement of the distal interphalangeal joint range with the finger in semiflexion. Figure 3—Measurement of the interphalangeal joint range of the thumb.

results obtained in 138 cases of flexor grafts in the fingers and thumb. At the Berne meeting of the Société Internationale de Chirurgie Orthopédique et de Traumatologie in 1954 a small group of surgeons studying these problems agreed to adopt Boyes's method of classification of results:

(*a*) *Flexor tendon action in a finger is recorded by measuring the distance by which the pulp of the finger tip fails to reach the distal crease of the palm* (Fig. 1).

Boyes however was not concerned with the restoration of profundus action alone in the presence of a normal sublimis tendon, and in this special group an additional measurement is necessary which calls for an explanation. The disability caused by loss of active flexion of the terminal phalanx is most noticeable in the index and middle fingers because these fingers take part in the thumb-finger grip. Controlled terminal flexion of these fingers is of importance to all individuals, but it is of special significance in all four fingers to those who need precision action. Replacement of the profundus tendon by free grafting will restore a useful flexion range to the terminal joint in most cases, but this range of movement tends to diminish as the finger is flexed into the hand.

Measurement of the finger tip to distal crease deficiency gives only a partial picture of the functional result, and it is instructive also to record the actual range of terminal interphalangeal joint movement when the finger is semiflexed (Fig. 2).

(*b*) *Flexor tendon action in the thumb is estimated by the range of active movement of the interphalangeal joint* (Fig. 3).

Following Boyes's method, the angle is expressed as a percentage of the passive range. Percentage of joint movement $= \frac{\text{voluntary flexion}}{\text{passive flexion}} \times 100$. The actual joint range is also presented as a check against this method of assessment.

SELECTION OF CASES

Flexor digitorum profundus—Operative treatment was restricted to those fingers in which passive movement was good and in which sensation of the finger tip was satisfactory, or could be restored by digital nerve suture. Arthrodesis was advised when these conditions were not satisfied. The total number of fingers operated upon was twenty-eight, of which six could not be traced. At least four of these cases are known to have been good so that their rejection from the series does not materially affect the conclusions.

Flexor digitorum profundus and sublimis—The operative indications in this group were less uniform. Grafting was not advised for tendon divisions in a single finger when the passive range was poor or when there was an irreparable complete nerve lesion. It was, and still is, considered that the better treatment in these cases is amputation. The index finger is an exception to this general rule and has always been considered a special case on account of the important thumb-index pinch. In the severely damaged hand with multiple flexor tendon divisions or loss of several digits, grafting has been performed under less favourable conditions on the grounds that even a small return of voluntary flexion would benefit the hand.

The total number of fingers treated was 109. It was not possible to trace sixteen cases but of this number at least half are known to have been satisfactory. A further three cases have been excluded because the fingers were in a poor condition and were in fact unsuitable for operation. There remain ninety fingers in which the end-results are known; in this group were eleven fingers in which grafting operations had been previously performed and had failed.

Flexor pollicis longus—Flexion of the interphalangeal joint of the thumb is essential for a good pinch grip. The normal range of flexion varies between 80 and 90 degrees, but an active range of 30 or 40 degrees is adequate for most purposes. The indications for grafting can therefore be less stringent than for the fingers; a moderate restriction of passive range does not contra-indicate operation.

The total number of cases was thirty-seven, all of which have been traced and are included in the series.

OPERATIVE TECHNIQUE

Exposure—Exposure has in all cases been made by a mid-lateral incision throughout the full length of the finger, the incision being deepened to the fibrous tendon sheath, passing posterior to the vessels and nerve. The palm has been opened by incisions placed in the appropriate crease lines. During the past few years a wider and more satisfactory exposure has been made by combining the finger and palm incisions and raising a large flap (Figs. 4 and 5). This incision owes its development to the influence of Rank and Wakefield (1952), although they prefer to carry the finger flap anterior to the neurovascular bundle. The middle and ring fingers do not so easily lend themselves to flap exposure and as a rule the limited approach is still employed. It is possible, however, by choosing suitable skin lines to link the two incisions, and by so doing the difficult area at the base of the finger can be fully exposed.

FIG. 4

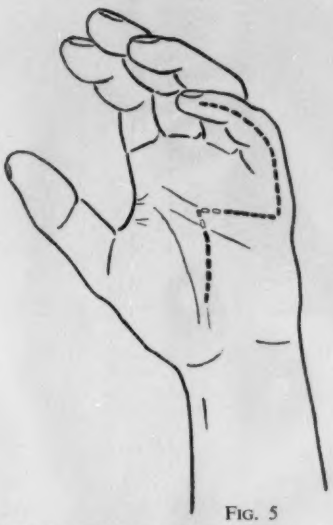


FIG. 5

FIG. 6

Figure 4—Incision for exposure of the index finger and palm. Figure 5—Incisions for exposure of the little finger and palm. The two incisions may be joined. Figure 6—Incisions for exposure of the thumb, palm and forearm. The proximal incision for removal of palmaris longus is also shown.

The thumb exposure requires three incisions—a mid-lateral incision on the radial side of the thumb, an incision the full length of the thenar crease and an angled incision above the wrist (Fig. 6). This approach has been used in all cases and has proved most satisfactory. It has occasionally been necessary to place the thumb incision on the ulnar side when the digital nerve on this side required repair. Figures 7, 8 and 9 show the exposure of the index finger, the little finger and the thumb.

Source of the graft—The grafts have been taken from palmaris longus, extensor digitorum longus to the fourth toe, and plantaris. Sublimis tendon was used in one case only before it was thought that a more slender graft would become revascularised more rapidly. It is interesting to note however that Kyle and Eyre-Brook (1954) have recently expressed their preference for sublimis. Flexor carpi ulnaris was used once for a special reason; the result was effective but some bow-stringing occurred owing to the difficulty of preserving satisfactory pulleys around the bulky tendon. In any case there is no need to sacrifice this important muscle. On four occasions an elongation of the flexor pollicis longus at its musculo-tendinous junction was performed and the tendon advanced to its insertion. The elongation is limited to one and a half inches and this restricts its use to tendon divisions in the distal part of the thumb. Theoretically the method is attractive, as it is probable that the blood supply of the tendon is not completely lost. The results in the four cases, although reasonable, did not reach the standard achieved by free grafting and the method was not continued. Elongation of the flexor digitorum profundus was used on one occasion and was successful, but this also has a limited application and is not recommended.

FIG. 7

Exposure of the index finger, showing the tendon graft, the slings and the digital nerve. The hand is held in a Fisk hand splint of sheet lead.

FIG. 8

FIG. 9

Figure 8—Exposure of the little finger, showing the tendon graft, the slings and the digital nerve, which has been sutured. The tension of the graft is demonstrated. Figure 9—Exposure of the thumb, palm and forearm, showing the tendon graft and its distal attachment by a Bunnell withdrawal suture. The tension of the graft is demonstrated.

The palmaris longus tendon, present in approximately 90 per cent of individuals, is usually of good quality and convenient size. Its length is only just sufficient and it cannot be used if extra length is needed for passing the graft through a tunnel in the terminal phalanx. It is the tendon of choice for the thumb because a wrist incision is part of the thumb exposure. Palmaris longus was originally removed by open dissection as it was thought essential to remove a plentiful covering of paratenon with the tendon. This method was abandoned when it was found that sufficient paratenon came away with the tendon when a limited exposure was made. The long forearm scar is unsightly in contrast with the two small scars which become almost invisible (Fig. 6).

The long extensor tendon of a toe is removed by open dissection, for it is difficult to extract it through small incisions (Fig. 10). It is of good quality and of ample length. Because of the short extensor muscle its loss does not seriously affect a toe, except for the little toe which does not possess a second extensor muscle. The skin wound may heal slowly unless complete haemostasis is secured and a careful skin suture performed. These tendons are the obvious choice when several grafts are required.

Plantaris, a very variable tendon, has its special value which deserves to be more widely known. It has the advantage of being thin and is the longest tendon in the body. It is the tendon of choice for the replacement of profundus in the presence of a normal sublimis because its slender character allows it to slip easily through the thecal canal leaving the sublimis undisturbed. Its presence cannot be determined until an exposure is made, but if it is absent no harm has been done and a toe extensor can be used. Two small incisions are needed, one at the medial side of the tendo Achillis and the other placed three finger breadths behind the medial border of the tibia in the mid-calf (Fig. 11). The tendon lies between the gastrocnemius and the soleus muscles.

FIG. 10

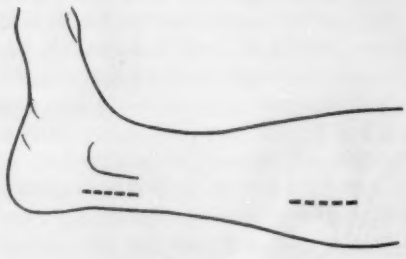


FIG. 11

Figure 10—The incision for removal of the toe extensor tendon. Figure 11—Incisions for removal of the plantaris tendon.

Apart from the special indications, the relative merits of these three tendons are difficult to assess and the choice is probably not one of great importance.

Suture of the graft—The suture material has been single-strand stainless steel wire (0·0046 of an inch) in all except three early cases in which ophthalmic silk was used. The wire has not been removed except in cases where a Bunnell withdrawal suture (Bunnell 1944) was used for the terminal fixation. In no case has any harm resulted from the permanent presence of the wire. The needles, which are swaged to the wire, are one inch long and are made in a very thin bayonet shape. They are constructed from a steel which will bend before it will break; this feature makes it possible to bend the needle into a suitable shape to negotiate awkward corners.

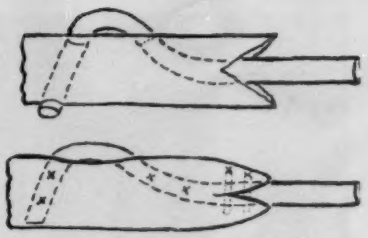


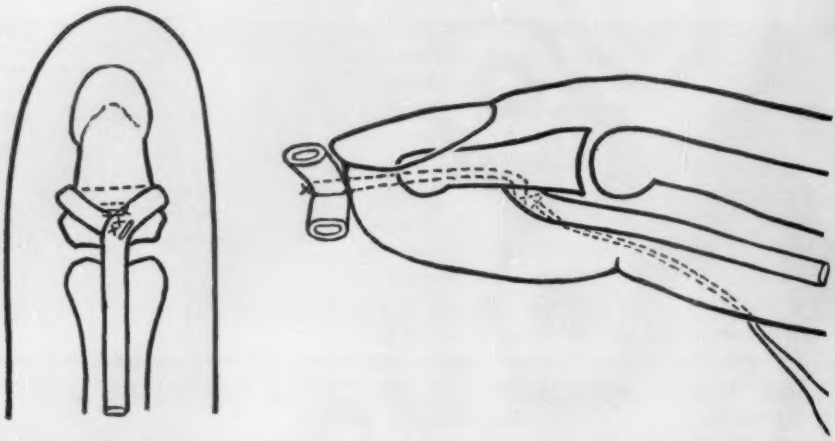
FIG. 12
Interlacing suture for the proximal junction of a slender graft.

The proximal suture has been performed by the well known Bunnell end-to-end stitch which is ideal when the tendons are of equal size, or by a particular form of interlacing stitch which is more suitable when tendon and graft are of different cross-sections (Fig. 12). As the majority of the grafts in this series were of the latter type, it became standard practice to use the interlacing suture. This junction has always been sited well proximal in the palm and the suture area has been covered by sewing lumbricalis muscle around it. In the case of the thumb, the suture is placed proximal to the wrist joint and covered by muscle if the graft is sufficiently long (Fig. 13).

Several methods of distal suture have been used—suture to the remaining tag of tendon, the Bunnell withdrawal suture, and looping the graft through a transverse hole in the terminal phalanx (Fig. 14). The first method was given up as it was thought to be less secure and more difficult to perform neatly. The Bunnell suture is a very neat and effective method and has been used when early movements have not been encouraged; it is the method of choice in children. Fixation of the graft through a transverse hole in the phalanx is also a tidy procedure and is, in my opinion, the most secure of all methods. The tunnel must be made in precisely the correct position through the broadest part of the bone. If it is placed too far dorsally there is a danger of injury to the nail root. The tendon is sutured to itself distal to the joint in order to avoid adhesions in front of the joint (Fig. 15).

FIG. 13
Exposure above the wrist to show the graft joined to flexor pollicis longus by an interlacing suture. This photograph was taken during the process of covering the junction by muscle. Flexor carpi radialis is retracted.

Tension of the graft—It is said that a tendon graft tends to shrink and that allowance should be made in judging the length of the graft. I am not convinced of the practical significance of this observation but there can be little doubt of another more important consideration. The muscle of the divided tendon shortens as it adapts itself to the inevitable retraction. It is not unreasonable to assume



(*a*) FIG. 14 (*b*)

Methods of distal fixation of the graft: (*a*) Graft passed through a tunnel in the distal phalanx and sutured to itself. (*b*) Bunnell withdrawal suture. The wire passes to the sides of the phalanx and not through the bone. A tightly rolled bolster of gauze is used in preference to a button.

that the muscle will return to normal or near normal length with the return of active use after a successful grafting operation, provided the period between injury and operation is not too long. It follows, therefore, that the tension under which the graft is placed should be a little greater than that which would appear to be normal at the time of operation. Normal tension under anaesthesia can easily be gauged by comparison with the other fingers, the index finger being flexed least and the little finger flexed most. In all the cases reported here the proximal suture has been performed first and the tension adjusted at the distal suture, the finger finally lying in a slightly more flexed position than would appear correct (Fig. 8). The same principle has been applied to the thumb (Fig. 9). It has been found in practice that a digit has rarely been put up too tight, but the opposite error has been made on several occasions. It is interesting to note that even in patients in whom there has been a long interval between injury and operation, it has rarely been found necessary to use another muscle as a motor. In 129 cases the average delay amounted to five and a half months; in the remaining twenty cases the delay varied between eighteen months and twenty-four years, and there were nine cases in which the interval was over three years. Contrary to expectations, there was no appreciable difference in the results of the short and long interval cases. One graft for profundus division with a four and a half years interval regained 30 degrees active range of the terminal interphalangeal joint. A graft for profundus and sublimis division after three years delay obtained a range of movement from full extension to half an inch from the distal palmar crease. A thumb graft regained 80 degrees range after four years (Fig. 16), and another obtained 70 degrees after ten years delay.

FIG. 15

Exposure of the finger to show a graft replacing the profundus tendon. The sublimis tendon is not disturbed. The graft is passed through a transverse tunnel in the phalanx.

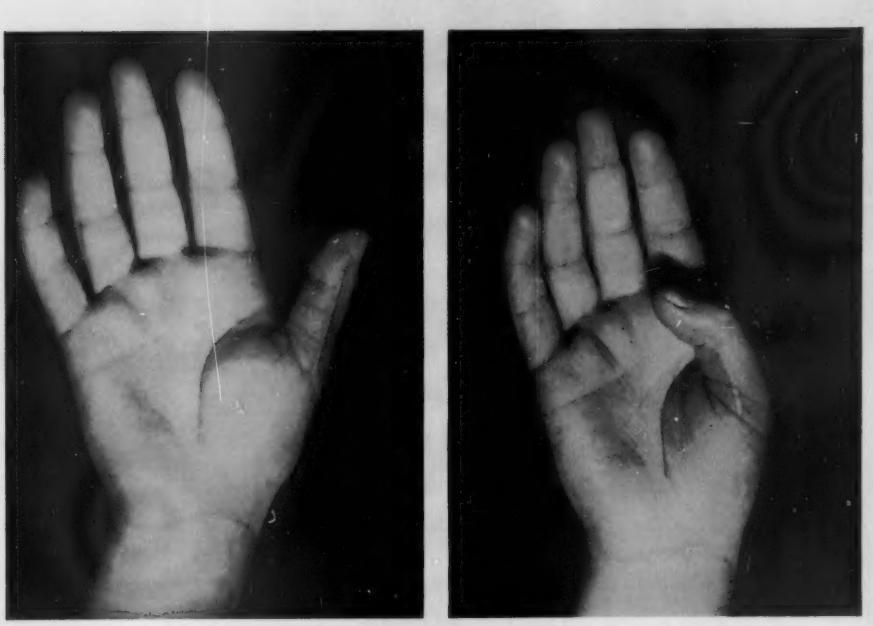


FIG. 16

The result obtained by a tendon graft to replace flexor pollicis longus, showing extension and flexion. The operation was performed four years after injury and these photographs were taken four months later. The flexion range is 40 degrees, which increased to 80 degrees eighteen months after operation. This is the same hand as is shown in Figure 9.

Treatment of the fibrous sheaths—The flexor tendons pass through a fibro-osseous canal in the fingers which holds the tendons against the flexor surface of the three finger joints. This fibrous tunnel or thecal sheath is lined by a synovial membrane which suspends the tendons in a mesentery. The theca is strengthened over the middle of the proximal and middle phalanges by transverse bands—the digital vaginal ligaments. It is our custom to remove the entire thecal sheath except for the vaginal ligaments, which are carefully retained as slings. When the theca has been severely damaged, or is so scarred that it needs to be completely removed (Fig. 17), it is necessary to form new slings to prevent bow-stringing of the graft (Fig. 18). These slings or pulleys may be constructed from a short length of graft.

FIG. 17

FIG. 18

Figure 17—Exposure of middle finger showing a previous failed graft. Note the very short graft. No thecal sheath remains. Figure 18—The same case, showing the new graft in place and the reconstructed tendon slings.

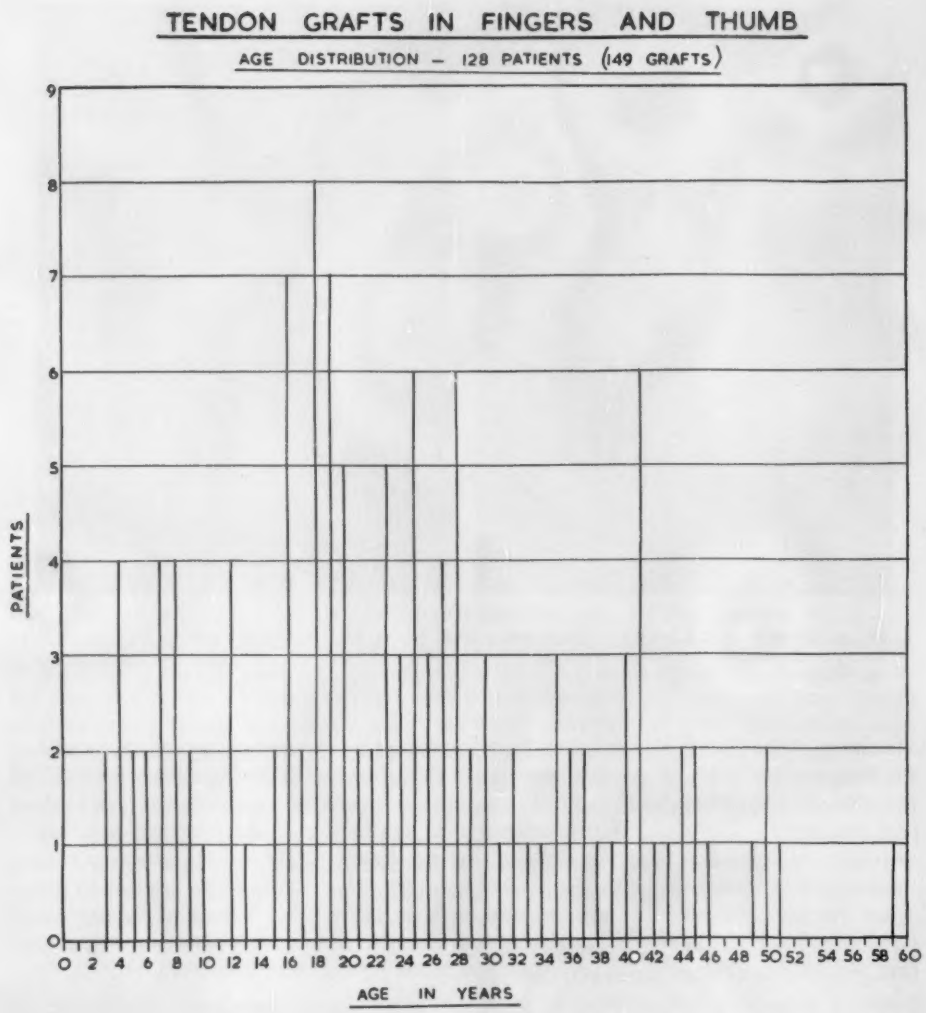

TENDON GRAFTS IN FINGERS AND THUMB
AGE DISTRIBUTION – 128 PATIENTS (149 GRAFTS)
PATIENTS
0 1 2 3 4 5 6 7 8 9
0 2 4 6 8 10 12 14 16 18 20 22 24 26 28 30 32 34 36 38 40 42 44 46 48 50 52 54 56 58 60
AGE IN YEARS

FIG. 19

The majority of injuries occurred in patients aged between sixteen and thirty years.

TENOLYSIS

The return of active movement after tendon grafting is often slow and much patience is needed. In those cases in which the flexion range ceases to improve after six to nine months and remains obstinately less than the passive movement, it is probable that the tendon flow is limited by adhesions. It is in these cases that a considerable improvement can sometimes be obtained by tenolysis, and failure changed to success.

The operation is extensive and needs to be performed with meticulous care. It is indeed a more difficult procedure than a tendon graft. A complete exposure is essential, for the full length of the graft must be freed from all its adhesions by gentle sharp dissection. Care is taken to preserve the pulleys, and, if this proves to be impossible, new pulleys must be

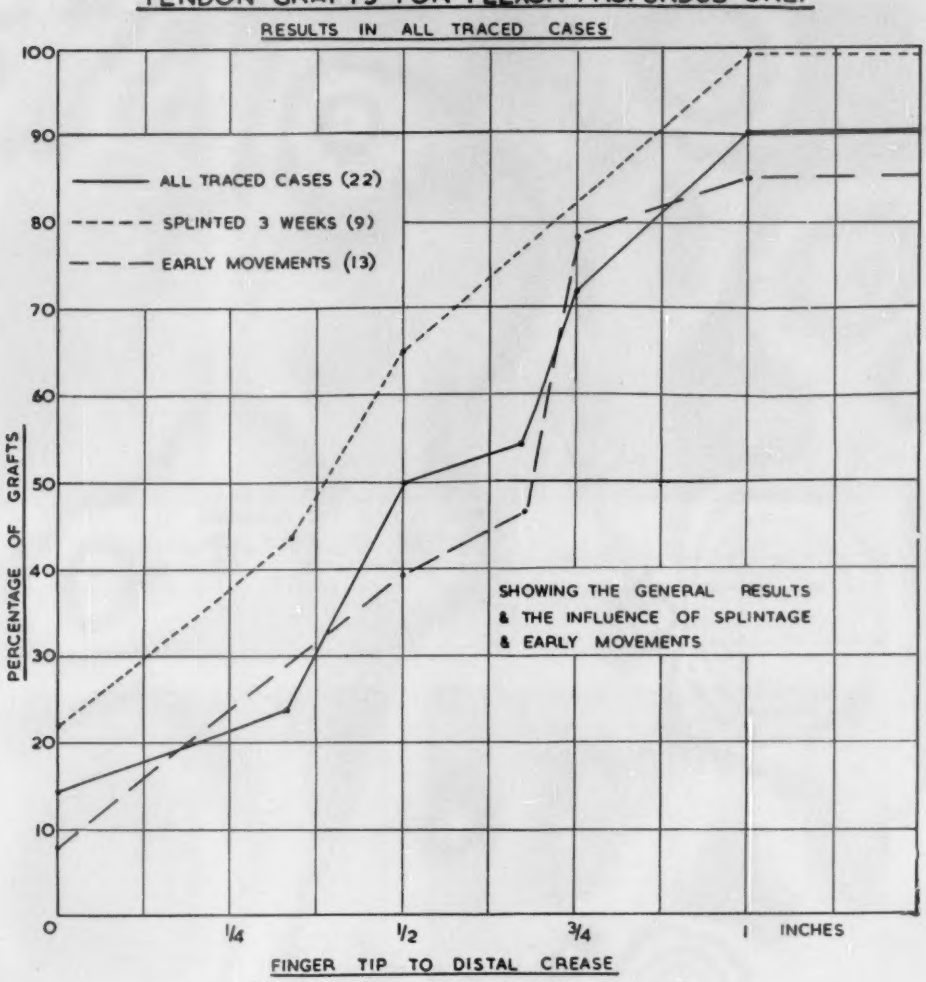


FIG. 20

The results obtained in all traced cases (twenty-two of a total number of twenty-eight), showing the distance by which the finger tip failed to reach the distal palmar crease. Note the influence of splintage and early movements.

constituted. The operation is not completed until it is clearly established that the finger will flex to the full passive range by pulling upon the proximal end of the graft.

Active movements are encouraged after a few days. During the past year cortisone has been given for ten to fourteen days after operation in an effort to limit adhesion formation (Carstam 1953). The number of cases so treated is yet too small to enable a fair judgment to be made, but the improved results suggest that cortisone may be beneficial.

Tenolysis has not been universally successful, but in no instance has the function been decreased and in several cases there has been a marked improvement.

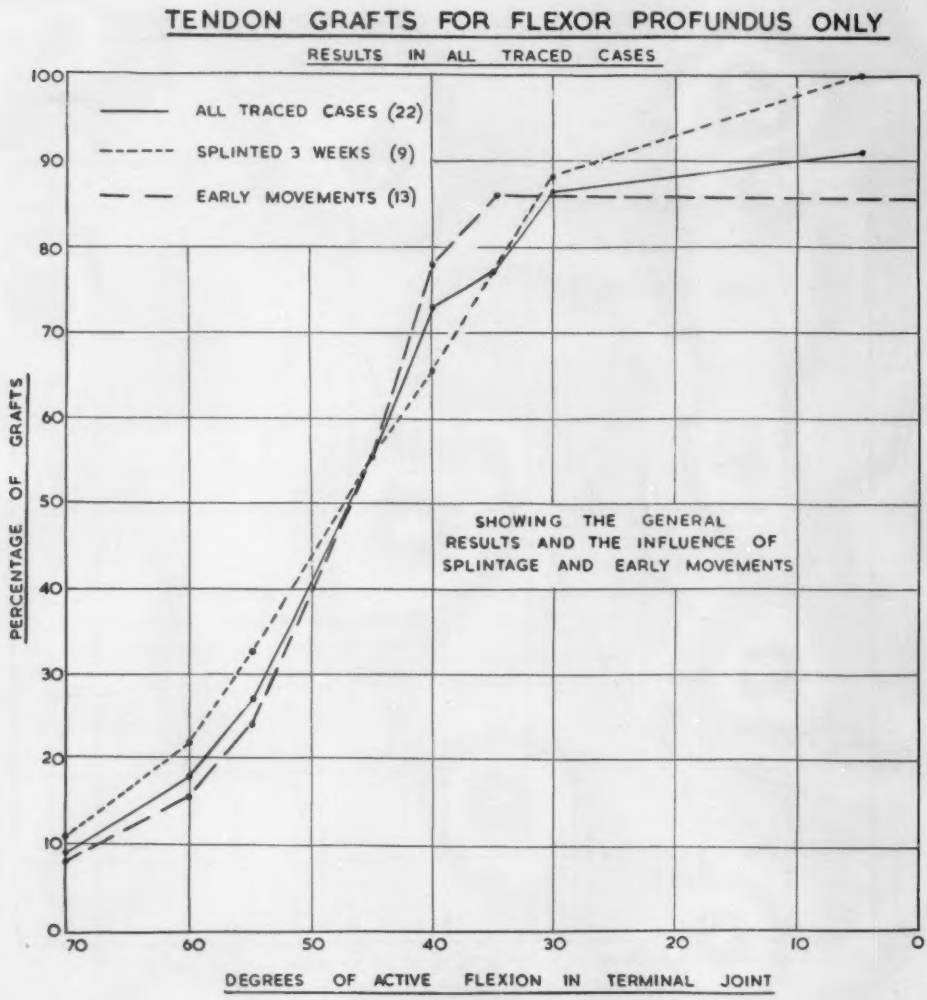


FIG. 21

The results obtained in all traced cases, showing the range of active movement in the distal interphalangeal joint when the finger is semiflexed. Note the influence of splintage and early movements.

RESULTS

Flexor profundus—The results are shown as graphs. The percentage of cases (individual digits) is plotted on the vertical axis. The horizontal axis shows: 1) the distance by which the finger tip fails to reach the distal palmar crease (Fig. 20); and 2) the active flexion of the terminal interphalangeal joint when the finger is semiflexed (Fig. 21). A reading of the first graph shows, for instance, that in 50 per cent of cases the finger tip reached to half an inch or less of the distal crease, and in 14 per cent of cases the tip touched the crease. The second graph shows that 86 per cent of cases obtained 30 degrees of active flexion and 9 per cent attained 70 degrees.

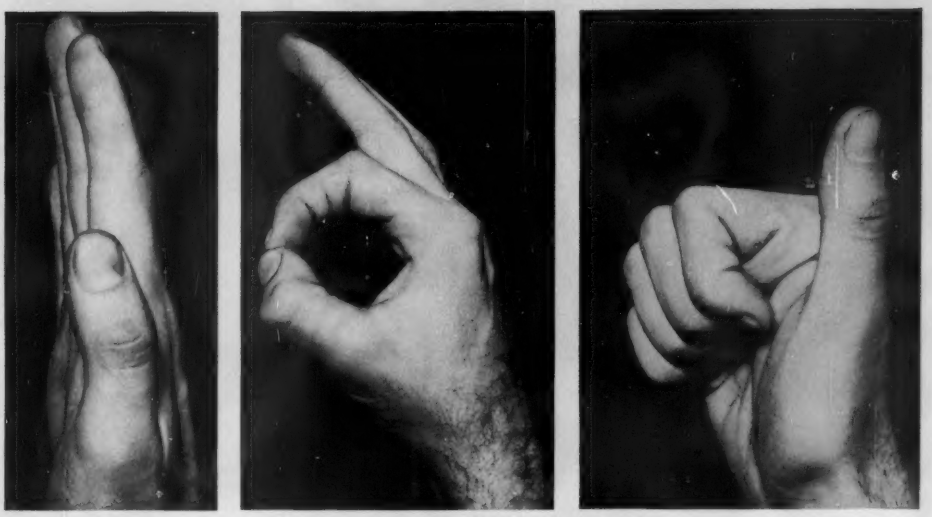


FIG. 22

The result obtained from a plantaris graft to replace the divided flexor profundus tendon of the index finger. Flexor sublimis was uninjured and was not disturbed. A digital nerve suture was performed at the same time. Terminal fixation was by a tunnel in the phalanx. Splintage was maintained for three weeks. The finger tip reaches to within half an inch of the distal palmar crease.

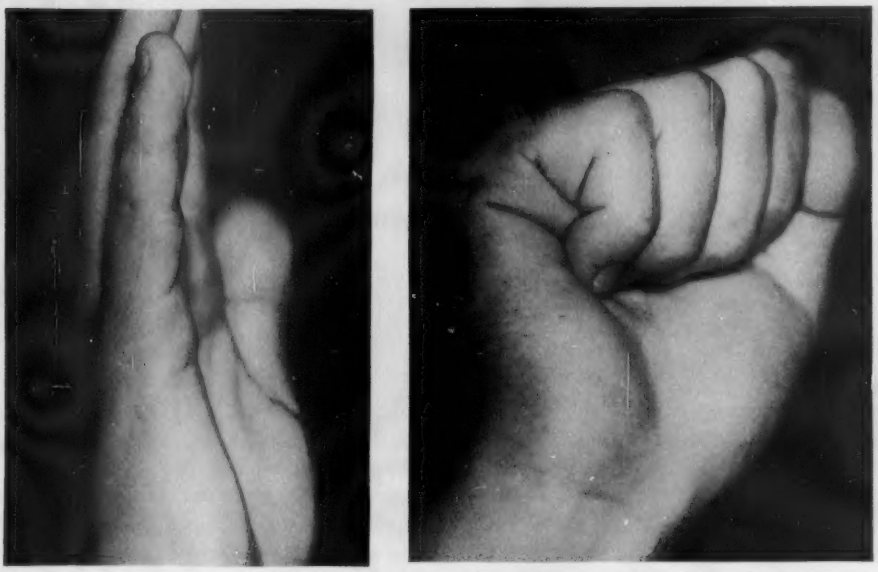


FIG. 23

The result obtained by a toe extensor graft to replace a divided flexor profundus tendon of the little finger. Flexor sublimis was uninjured and was not disturbed. Terminal fixation was by a withdrawal suture. Splintage for three weeks. The finger tip reaches the distal palmar crease.

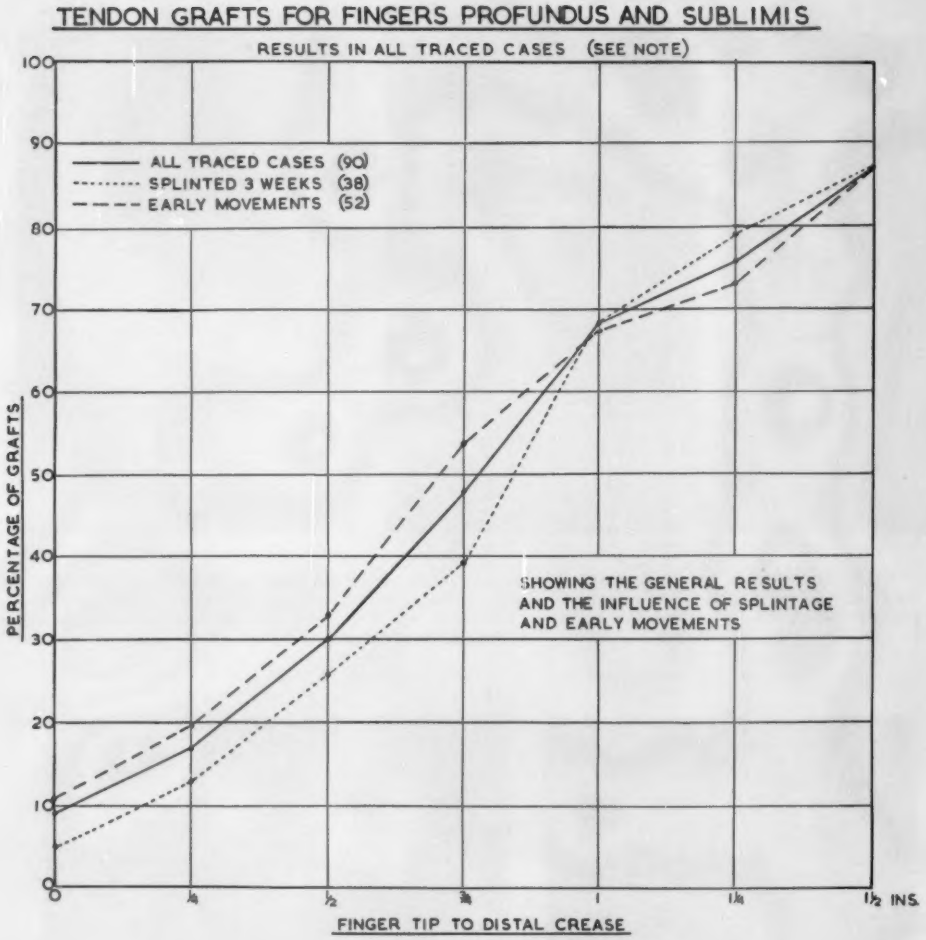


FIG. 24

The results obtained in all traced cases (ninety of a total number of 109), except for three cases which have been deliberately excluded because the fingers were in poor pre-operative condition. Two cases had complete nerve lesions and one had a previously failed graft with severe contracture. Note the influence of splintage and early movements.

Extension was full in all cases except four, in which there were flexion contractures at the terminal interphalangeal joints of 5, 5, 10 and 20 degrees.

There were three failures. In two instances the flexion range was very poor and for one of these patients an arthrodesis was performed; the other patient expressed herself as satisfied with the result. The third case failed owing to rupture of the graft at two weeks; the operation was repeated and 30 degrees of active flexion were obtained.

There were no complications and in no instance was a finger made worse by operation. This fact is of particular importance in this group, since an operation of this magnitude for a comparatively minor disability would stand condemned if it led to disaster.

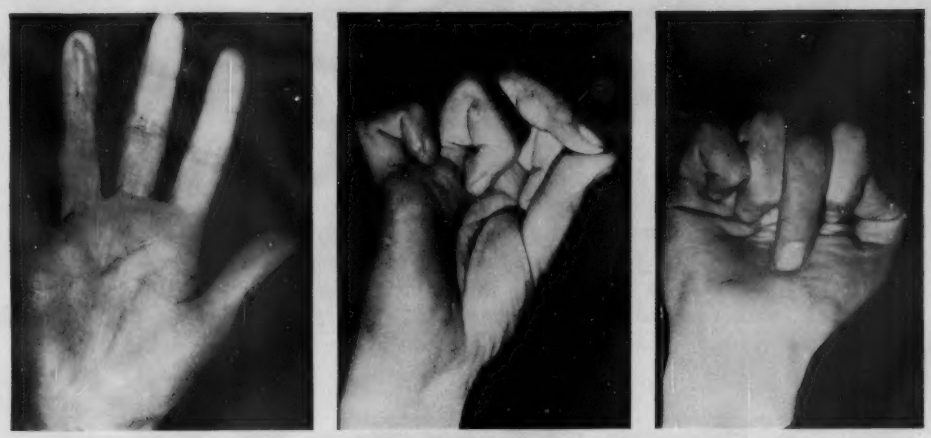


FIG. 25

The result obtained from a plantaris graft to replace divided flexor sublimis and profundus tendons of the middle finger. Terminal fixation was by a tunnel in the phalanx. Early movements. The finger tip reaches to within three-quarters of an inch of the distal palmar crease.

FIG. 26

The result obtained by a palmaris graft to replace divided flexor sublimis and profundus tendons of the middle finger. Terminal fixation was by a withdrawal suture. Splintage for three weeks. The finger tip reaches to within a third of an inch of the distal palmar crease.

The general inference to be drawn from these results is that replacement of profundus in the presence of a normal sublimis is justifiable if the state of the finger is satisfactory. It should be explained to the patient that there is a good expectation of restoring a reasonable flexion range, but that only if he is very fortunate will he recover full joint movement when the finger is fully flexed (Figs. 22 and 23).

Flexor profundus and sublimis—The results are presented in a similar manner showing the distance by which the finger tip fails to reach the distal palmar crease (Fig. 24). In this series of ninety grafts nearly 70 per cent of the fingers could reach to within one inch or less of the distal palmar crease, but only 9 per cent regained perfect flexion. There were twenty-nine cases (32 per cent) in which there was a flexion contracture at the proximal interphalangeal joint of over 5 degrees, and of these, twelve suffered from a contracture of 30 degrees or more. A flexion deformity of 30 degrees is not a troublesome disability although it may detract from the appearance of the hand when it occurs in the middle or ring finger. A deformity of more than 30 degrees cannot be regarded with complacency, although all these patients expressed themselves well satisfied with the functional improvement they had obtained. Contractures of this degree are disquieting. The reason for their development is obscure, for although some occurred in fingers already somewhat contracted, or in patients with multiple finger injuries, a few occurred in fingers which were in a perfect pre-operative state.

(*a*) (*b*) (*c*)

FIG. 27

The result obtained by a toe extensor graft to replace divided flexor sublimis and profundus tendons of the middle, ring and little fingers at one operation. The pre-operative (*a*) and post-operative (*b*) and (*c*) conditions are illustrated. Terminal fixation was by a tunnel in the phalanx. Splintage for three weeks. The finger tips reach to one and three-quarters, one and a half and one inch from the distal palmar crease. There is a flexion contracture of 20 degrees at the proximal interphalangeal joints.

Flexion contractures (eight cases) occurred more often when early movements were given; only four splinted cases developed contractures of over 30 degrees.

There were nine complete failures, six of which were due to adhesions and three to rupture of the graft without undue violence. One rupture occurred in a patient who had suffered from an original attrition rupture of his flexor tendon due to an arthritis of the wrist joint and it is probable that the same cause was responsible for the graft rupture. The three cases of rupture occurred in patients who had commenced gentle movements one week after operation. Two of the failures due to adhesions happened in patients who were treated by early movements and four in whom splintage was maintained for three weeks.

There were no complications apart from one instance of mild sepsis near the finger tip which rapidly responded to treatment. The final result was satisfactory.

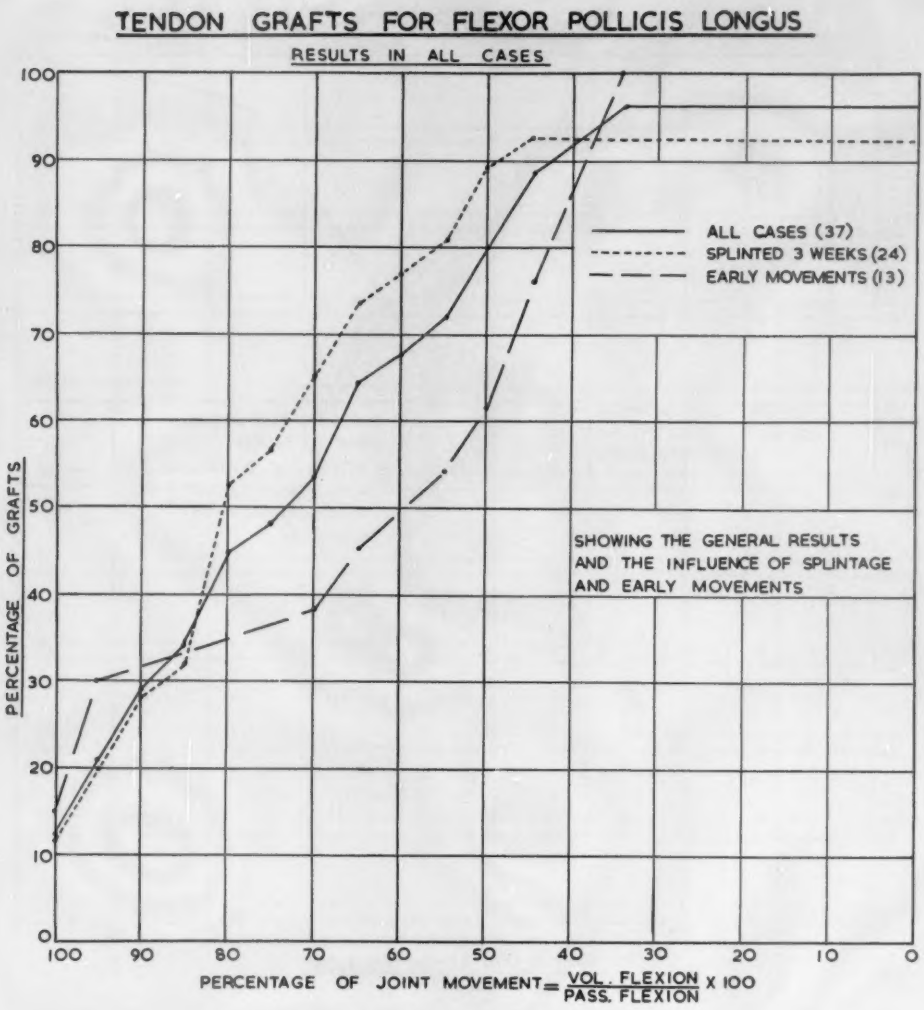


FIG. 28

The results obtained in all cases, showing the percentage of active joint movement in relation to passive movement. Note the influence of splintage and of early movement.

It seems reasonable to conclude that a satisfactory functional result may be expected in approximately 70 to 80 per cent of patients in whom the pre-operative state of the finger is good (Figs. 25 to 27).

Flexor pollicis longus—The results are shown in three graphs depicting: 1) the percentage of active joint movement in relation to passive movement (Fig. 28); 2) the active joint range measured in degrees (Fig. 29); and 3) the influence of the graft source upon the result (Fig. 30).

The general inference is that there is a high measure of success (Figs. 31 and 32). Eighty-five per cent attained 30 degrees of joint movement and 21 per cent reached 70 degrees (Fig. 29).

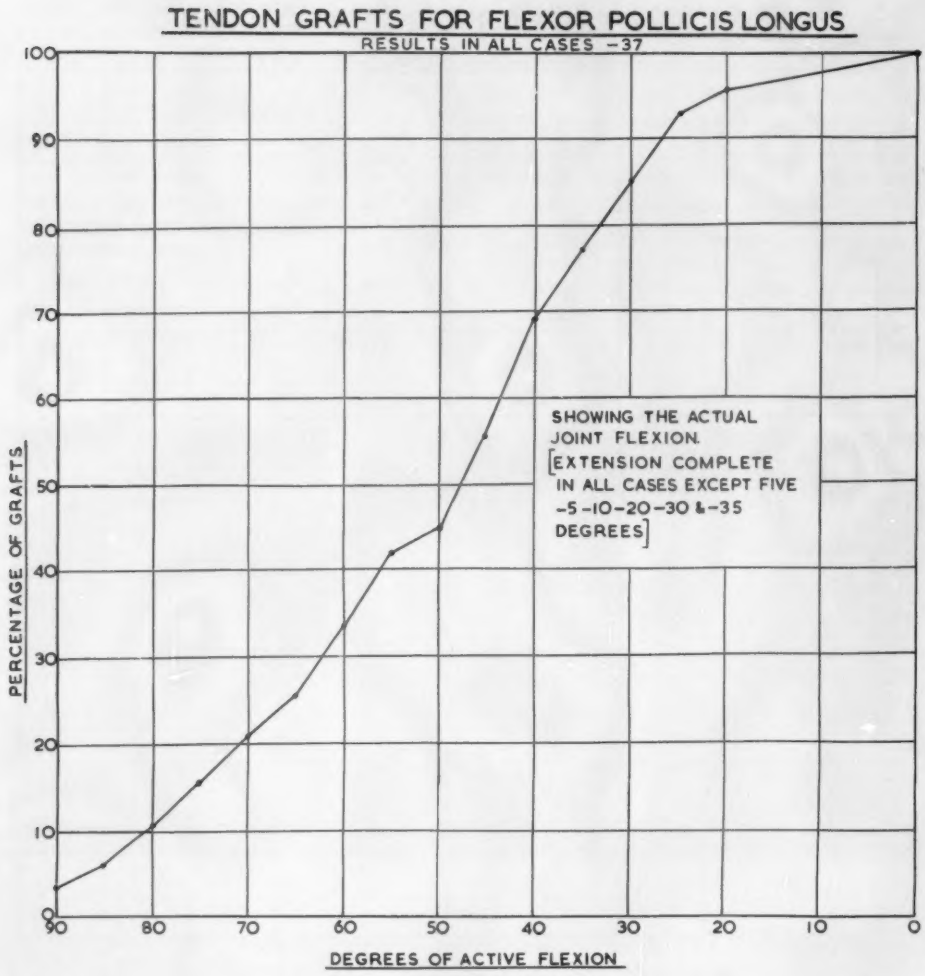


FIG. 29

The results obtained in all cases, showing the true joint range obtained.

In only one case did the graft fail—this was due to rupture two months later owing to unexpected severe strain. The patient was lifting a heavy suitcase from a railway carriage rack when it slipped and caught his thumb. The operation was repeated and he gained 45 degrees of active flexion.

Influence of the graft source—Palmaris longus and the toe extensor have been most frequently used and the results obtained from these grafts are compared. The thumb has been chosen as the conditions have been more uniform than in the finger. It will be seen that there is no essential difference in value, which confirms the impression gained during the years that the choice may be made for convenience and for the reasons already stated.

Influence of the after-treatment—It has been my opinion that gentle active movement commenced within a few days or a week of the operation gave more impressive results than were obtained

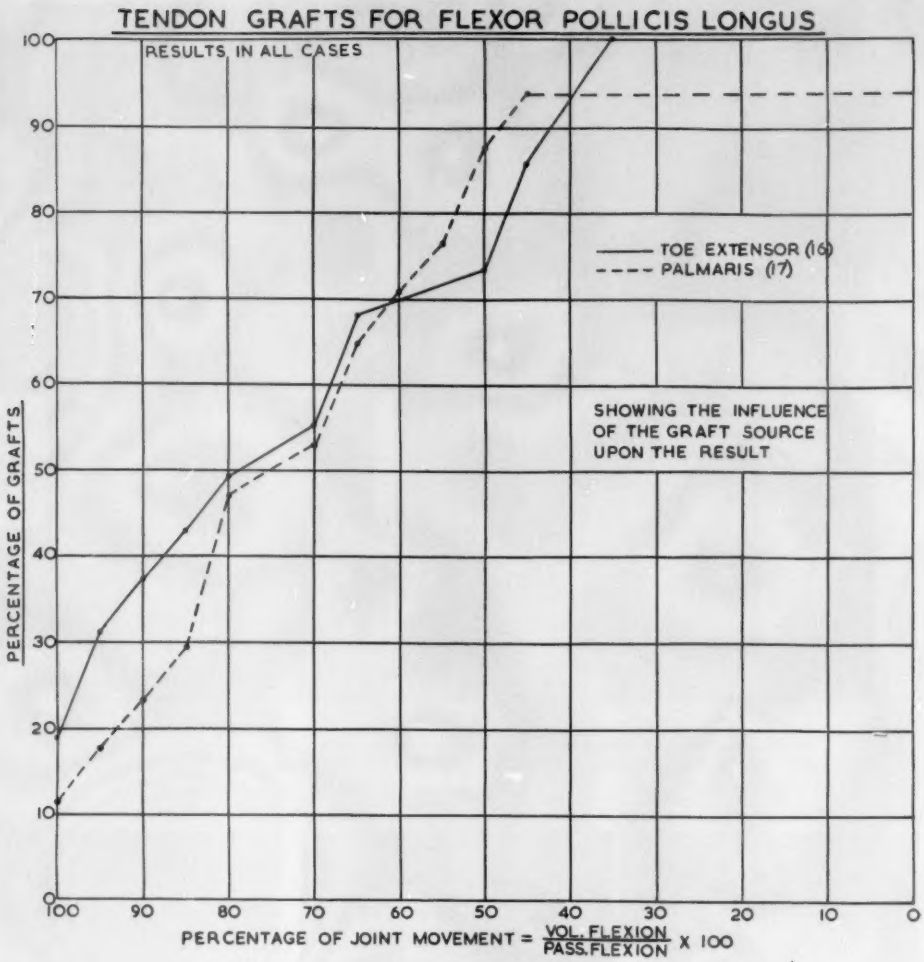


FIG. 30

The results obtained in thirty-three cases, showing the percentage of active joint range in relation to passive movement. The graph demonstrates the influence of the graft source. Four cases of elongation of the flexor pollicis longus at its musculo-tendinous junction have been excluded.

if splintage for three weeks was used (Pulvertaft 1948). It was therefore with great personal interest that these results were analysed. In each of the three series the influence of splintage and early movements has been compared (Figs. 21, 24 and 28), and it came as a surprise to find that there was very little difference (Figs. 33 and 34). It is noteworthy that Iselin (1955) has expressed the same opinion.

When splintage is employed it would seem wiser to hold the finger or thumb in considerable flexion. It is easier to overcome adhesions in flexion during the first few weeks following the release of splintage than to improve the flexion range. The judicious use of elastic traction will assist the return of extension, but there is no effective way of improving flexion apart from the patient's own active use.

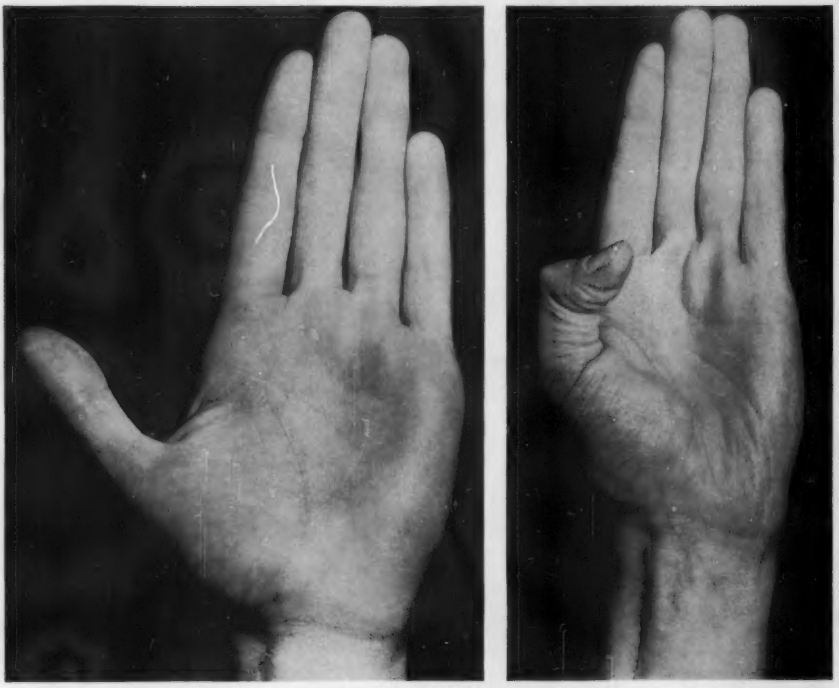
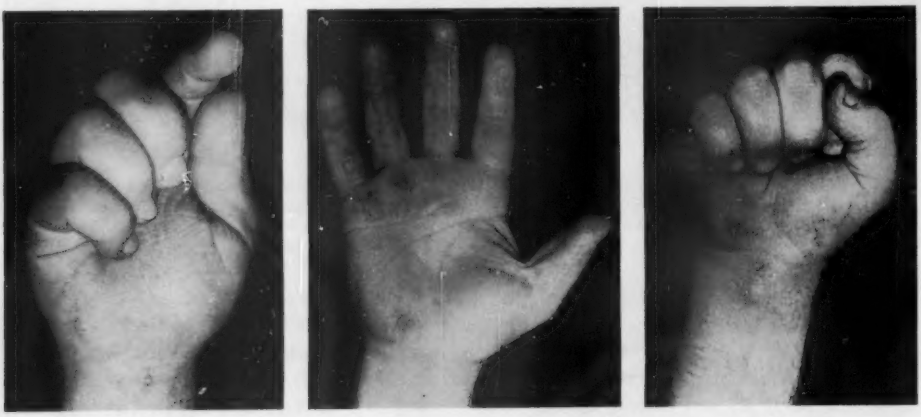


FIG. 31

The result obtained from a palmaris graft to replace a divided flexor pollicis longus. Terminal fixation was by a withdrawal suture. Splintage for three weeks. The interphalangeal joint range was 84 degrees.



(*a*) (*b*) (*c*)

FIG. 32

The result obtained by: 1) a palmaris graft to replace a divided flexor pollicis longus; 2) a short graft in the palm to bridge flexor profundus of the index finger, and local removal of the divided sublimis; 3) suture of the median nerve, all at one operation. The pre-operative (*a*) and post-operative (*b*) and (*c*) conditions are illustrated. Terminal fixation was by a withdrawal suture. Splintage for three weeks. The interphalangeal joint range is 55 degrees. The index finger has a complete range of movement.

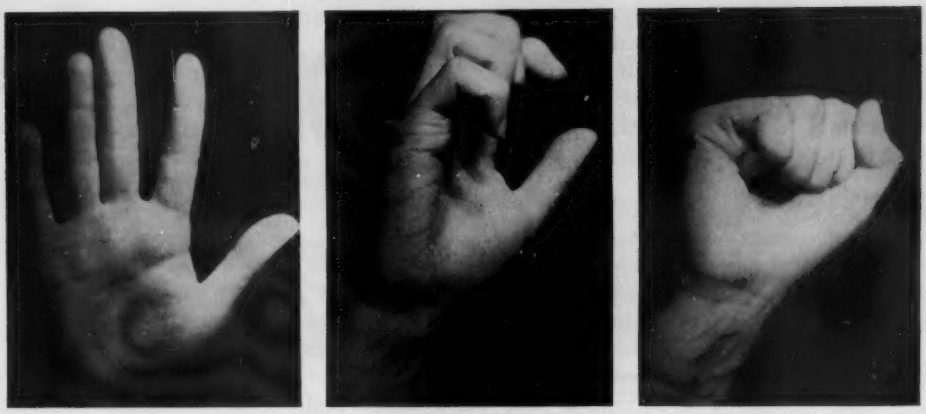


FIG. 33

The result obtained from a toe extensor graft to replace divided flexor sublimis and profundus of the little finger. Terminal fixation was by suture to tendon. Splintage for three weeks. Extension, semiflexion and full flexion are illustrated. The finger tip reaches the distal palmar crease.

FIG. 34

The result obtained from a toe extensor graft to replace divided flexor sublimis and profundus of the little finger. Terminal fixation was by suture to tendon. Early movements. Extension, semiflexion and full flexion are illustrated. The finger tip reaches the distal palmar crease.

SUMMARY

1. A series of 149 consecutive cases of flexor tendon grafting in the fingers and thumb is reviewed and the results are analysed. A description of the technique which has evolved from this experience is given.

2. Replacement of a divided flexor digitorum profundus in the presence of an intact sublimis tendon restored a useful range of movement in 80 per cent of cases.

3. Replacement of both flexor digitorum profundus and sublimis gave good results in 70 to 80 per cent of cases.

4. Replacement of a divided flexor pollicis longus tendon gave good results in 85 per cent of cases.

5. The particular tendon used for the graft did not materially influence the result, but there are special indications for the use of the different tendons.

6. The choice of splintage or of early movement after operation does not appear to have a significant bearing upon the result.

7. The most important factor in determining the result is probably a precise and gentle surgical technique and complete haemostasis. Minor differences in method are of little importance. The failures in this series were due more to faulty performance of the operation than to any other single factor.

I would like to thank the many surgeons who have helped me with these cases, with special mention of Douglas Reid and Konstantin Velaskakis. I am much indebted also to Miss Peggy Coupland and to Mr J. S. Fayers, both of the Derbyshire Royal Infirmary, for their able assistance with the secretarial and with the photographic work.

Figures 7, 22 and 33 are reproduced from the Proceedings of the Sixth Congress of the International Society of Orthopaedic Surgery and Traumatology.

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SUDECK'S ATROPHY IN THE HAND

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Sudeck's atrophy, or post-traumatic osteodystrophy, is considered by most surgeons in Great Britain to be a relatively rare condition, although in Europe and North America it is claimed by some to be the most common cause of prolonged disability after injury (Jordan 1940, Gurd 1938, Scheibe and Karitzky 1954, Herrmann, Reineke and Caldwell 1942). Only one British contribution appears in the extensive world literature on the subject (Middleton 1937), and Watson-Jones considers it to be a severe form of disuse atrophy (Watson-Jones 1952).

Much has been written about the treatment of this condition, particularly in relation to surgical attacks on the sympathetic system (Brücke 1946, Evans 1946, Heyman 1924, Herrmann 1935, Toumey 1948, Kiær 1948), parasympathetic stimulation (Middleton 1937), the application of plaster splints, x-ray therapy (Mumford 1938), physical therapy (Sudeck 1938, Noble and Hauser 1926, Jordan 1940), and more recently on the use of vasomotor relaxants and cortisone (Dwyer 1952, Rose 1953, Roland 1952, Birkenfeld 1954, Marti 1954).

It is the purpose of this paper to show firstly that Sudeck's syndrome of the acute type is not uncommon in the hand and, secondly, that it responds to conservative treatment if the diagnosis is made within six weeks of the onset of symptoms. No attempt will be made to add yet another theory to explain the etiology, but some observations on the occurrence of thickening of the palmar fascia in association with Sudeck's syndrome will be recorded.

TABLE I

CAUSATIVE FACTORS IN THIRTY-SEVEN CASES OF SUDECK'S ATROPHY

Colles's fracture	16 (14 women)
Smith's fracture	5 (4 women)
Fractures about the wrist:	
Scaphoid	1
Triquetral	1
Metacarpal	2
Finger injuries:	
Crush	2
Laceration of pulp	1
Crush injuries of hand and wrist (without fracture)	3
Brachial plexus injuries (one with dislocated shoulder)	2
Arm injuries:	
Dislocation of elbow	1
Fracture of humerus	1
Hemiplegia	1
Burn of forearm and hand	1

INCIDENCE AND CLINICAL FEATURES

During the three years 1952–54, 80,000 new patients were seen in the Accident Services of the Luton and Hitchin Group of hospitals, and in this period Sudeck's syndrome was recognised on thirty-seven occasions. Two-thirds of these cases were in women, and the

FIG. 1

An example of Sudeck's syndrome three weeks after the onset of symptoms. Swelling and colour changes are accompanied by partial obliteration of skin creases and loss of Lange's lines. The fingers are fixed in flexion and already show tapering The swelling in this case is more than usual (120 cubic centimetres).

FIG. 2

Typical spotty appearance of Sudeck's atrophy involving carpus, metacarpals and phalanges. The shafts of the metacarpals and phalanges are only slightly affected.

average age of the whole group was fifty-eight years (youngest thirty-three, oldest seventy-six). The causative factor was most frequently an injury in the region of the wrist (Table I) (Bethge and Meyer 1953).

The symptoms of pain, swelling and dysfunction developed at an average of eight weeks after injury and were accompanied by the following signs: hyperaemia, loss of Lange's lines, obliteration of skin creases, loss of tissue turgor in the pulp, changes in skin temperature and colour, nail ridges, limitation of movement and tenderness. Power of grip was grossly impaired (average 10 per cent of normal) and the volume of the hand increased by 10–20 per cent.

The pain is burning or aching in character and several patients described it as being deep. It is not relieved by splinting or by short-wave diathermy. The swelling is not due entirely to oedema. When the hand volume is increased by as much as 100 cubic centimetres pitting on pressure can be demonstrated, but when this oedema has responded to treatment there is left a visible and palpable thickening of the soft tissues. The degree of obliteration of the

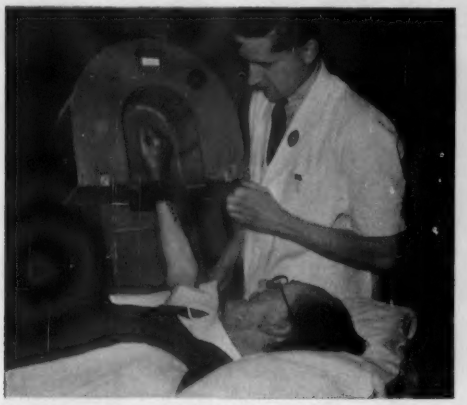

FIG. 3

FIG. 4

Figure 3—The physiotherapist is showing the patient how to exercise the intrinsic muscles while the hand is elevated and covered with wax and polythene in a warm cabinet. Figure 4—The operating lever of this drilling machine is surrounded by a Perspex cabinet warmed by hot air. The hand works above shoulder level.

skin creases opposite the joints corresponds with the amount of swelling, but the loss of Lange's lines is not related to the swelling of the soft tissues. Even when the volume of the hand returns to normal, Lange's lines remain less marked for many months.

The hand is held immobile with the fingers in a position of semiflexion (Fig. 1), and attempts at movement result in only a small range at the metacarpo-phalangeal joints. The terminal pulps of the fingers are tapered and feel less resilient than normal, and the return of the skin colour after pressure is brisk. Transverse ridges appear on the nails. In the early stages the hand is deep pink and the skin feels warm. Later, the skin takes on a mottled cyanotic appearance and feels cold and dry.

Radiographic appearance—The characteristic radiographic appearance is not present in the early stages, but takes six to eight weeks to show, and it is for this reason that the diagnosis is often overlooked. The spotty rarefaction that involves the ends of the phalanges and metacarpals is unmistakable (Fig. 2). Sometimes the carpus and the distal ends of the radius and ulna are involved, whereas the shafts of the bones are only slightly affected. The spotty appearance is quite unlike the generalised ground-glass appearance seen in disuse atrophy, which takes a much longer time to develop. Sometimes the clinical signs and symptoms have disappeared by the time radiographic changes can be seen. On occasions pain and dysfunction in more proximal joints follow (elbow and shoulder); here again, the same radiographic appearance can be seen but not until six weeks after the onset of symptoms in these joints.

In 1900 Sudeck described these typical radiographic appearances and considered them to be abnormal. More recently he ascribed them to an exaggerated but normal process of inflammation and repair (Sudeck 1938). It is true that slight radiographic changes can be seen in a small proportion of hands after injury without the signs and symptoms of Sudeck's syndrome. In a recent analysis of one hundred patients over the age of fifty with wrist fractures, radiographs taken eight weeks after the removal of plaster showed slight subchondral spotty osteoporosis in six cases. Well marked radiographic changes were always accompanied by signs and symptoms of Sudeck's syndrome.

TREATMENT

Treatment in our clinic is entirely conservative and consists in the application of heat, elevation of the limb, and graded function. We find that hot wax is just as effective in the relief of pain as blocking the cervical sympathetic chain with procaine. Elevation reduces the oedema by a measurable amount. Gradually increased function in conjunction with heat and elevation results in the relief of symptoms in every early case in an average time of six weeks. During the last four years the technique of applying hot wax, elevation and exercise therapy has been improved and is now standardised.

FIG. 5

This simple hand volume-measuring equipment is accurate to within 1 per cent because the hand and fingers are always trapped in a constant position.

The hand is immersed in hot molten wax for a minute or so until the hand feels warm and is then covered with a polythene glove. The limb is then elevated on an inclined plane with a completely enclosed radiant heat cradle to keep the hand temperature between 105 and 110 degrees Fahrenheit (Fig. 3). Exercises for the intrinsic muscles are then taught and supervised by the physiotherapist. This procedure is carried out twice a day for half an hour each time. The rest of the day is spent in the remedial workshop. The patient sits on a low chair and operates a machine with the operating lever raised so that the affected hand is kept above the level of the shoulder. Warmed air is blown into a transparent cabinet which surrounds the hand and maintains a temperature of 105 degrees Fahrenheit (Fig. 4). For the first week or two simple analgesics are necessary but they can be nearly always discontinued by the third week.

Each day the power of grip is recorded and the volume of the hand is measured with equipment designed by M. Eccles, a therapist on our staff (Fig. 5). This volume-measuring equipment is accurate to within 1 per cent and is based on the principle of water displacement.

Two case histories will serve to illustrate the salient features of Sudeck's syndrome.

CASE REPORTS

Case 1—A woman of seventy crushed the tip of her right ring finger in a folding chair. One stitch was required to close the laceration of the pulp and the wound healed in ten days, with some puckering of the scar. A month later she developed pain in the ring finger and in the two adjacent fingers. The pain gradually became worse and the three fingers stiffened. When she began to lose sleep she came

to hospital ten weeks after the injury. The three ulnar fingers were deep pink in colour, swollen and almost completely rigid. The skin creases opposite the joints were partly obliterated and Lange's lines were barely visible. There was a flame-coloured staining under the distal part of the nail of the ring finger and the healed laceration of the pulp was slightly tender. The three ulnar fingers were dry and cold. Radiographs showed slight spotty changes in the subchondral area of all the small joints of the ulnar three fingers. A month later there was a well marked Sudeck's atrophy of the ulnar side of the hand (Fig. 6). Volume of hand (Fig. 7) and power of grip (Fig. 8) returned to normal

FIG. 6

Case 1—There is a well marked osteoporosis of the terminal phalanx of the ring finger. Sudeck's changes are apparent in the three ulnar fingers and their metacarpals. The thumb and index finger are unaffected.

after three months' intensive treatment, and at this stage the physiotherapist noticed some thickening of the palmar fascia on the ulnar side of the palm. Six months later the hand was normal in all respects, except for a well marked Dupuytren's contracture on the ulnar side of the hand with dimpling of the skin of the palm, but with only slight limitation of extension of the ring and little fingers.

Case 2—A boxmaker aged fifty-four sustained a Smith's fracture which was reduced and splinted two hours after injury. The patient went back to modified work while in plaster, which was kept on for six weeks. Three days after the plaster was removed there was a sudden onset of pain, swelling and stiffness in the hand. It became deep pink in colour and function was lost. Lumps were found on the ulnar side of the palmar fascia. At the time of the first treatment the volume of the hand was 578 cubic centimetres (other hand 470 cubic centimetres) and the power of grip was 6 lb. (other hand 70 lb.) (Figs. 9 and 10). Intensive daily treatment for eight weeks resulted in a diminution of the volume of the hand to 454 cubic centimetres and an increase of power of grip to 58 lb., and the patient returned to normal work. Radiographs showed obvious changes (Fig. 11). The thickening of the palmar fascia was well marked a year after the onset of symptoms and there was no involvement of the other palm.

THE ASSOCIATION OF SUDECK'S ATROPHY WITH THICKENING OF THE PALMAR FASCIA

The thickening of the palmar fascia described in the above examples was first noticed by Eccles (1956) and it soon became apparent that it was a regular feature of this syndrome. In six cases there was palpable nodular thickening of the palmar fascia in both hands, in four

the condition was more evident on the side affected by Sudeck's syndrome, in fifteen others nodules were felt on the affected side only, and in three the thickening developed while the patient was under observation during treatment.

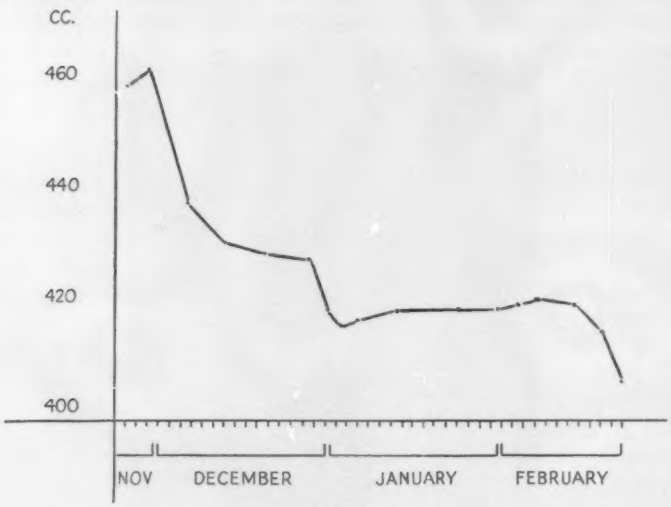

FIG. 7

Case 1—Volume chart of right hand. (Left hand 405 cubic centimetres.)

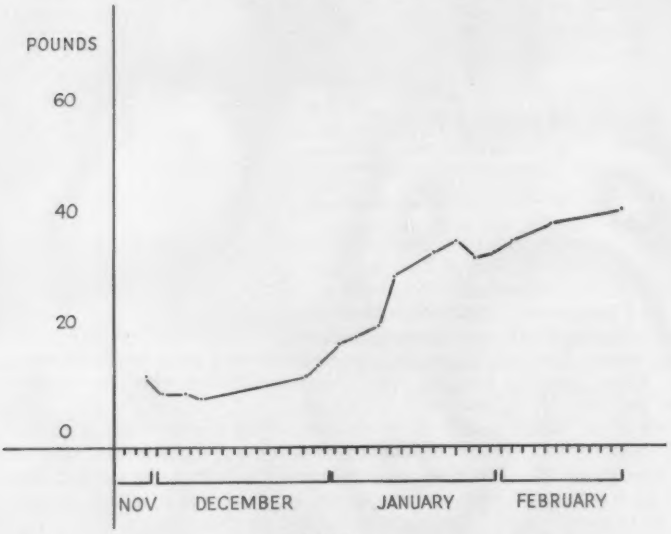

FIG. 8

Case 1—Power of grip chart, right hand. (Left hand 46 lb.)

Established Dupuytren's contracture (thickening of part of the palmar fascia with dimpling of the skin and deformity of at least one digit) was present before the onset of Sudeck's atrophy in three cases (bilateral in two), and in each of these the condition got worse for a time. One patient developed a contracture while under treatment, and four patients were noticed to

have contractures on the affected side at some stage in treatment, but no notes were made about the deformity when first seen. In only one patient of the total of thirty-seven was the palmar fascia not thickened or contracted.

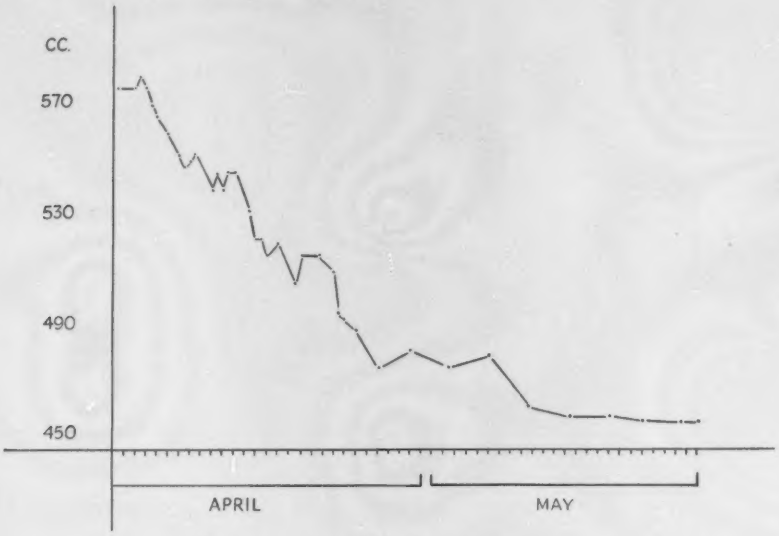

FIG. 9
Case 2—Volume chart of right hand. (Left hand 470 cubic centimetres.)

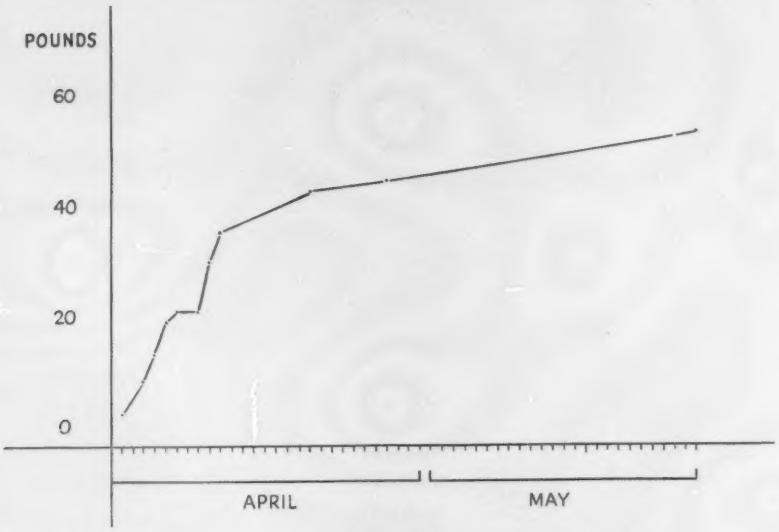

FIG. 10
Case 2—Power of grip chart, right hand. (Left hand 70 lb.)

From these observations it is clear that Sudeck's atrophy is nearly always accompanied by an alteration in the palmar fascia which may go on to a typical Dupuytren's contracture. These two conditions apparently have nothing in common except that they affect persons of about the same age. The fact that most of the cases reported in this paper occurred in women,

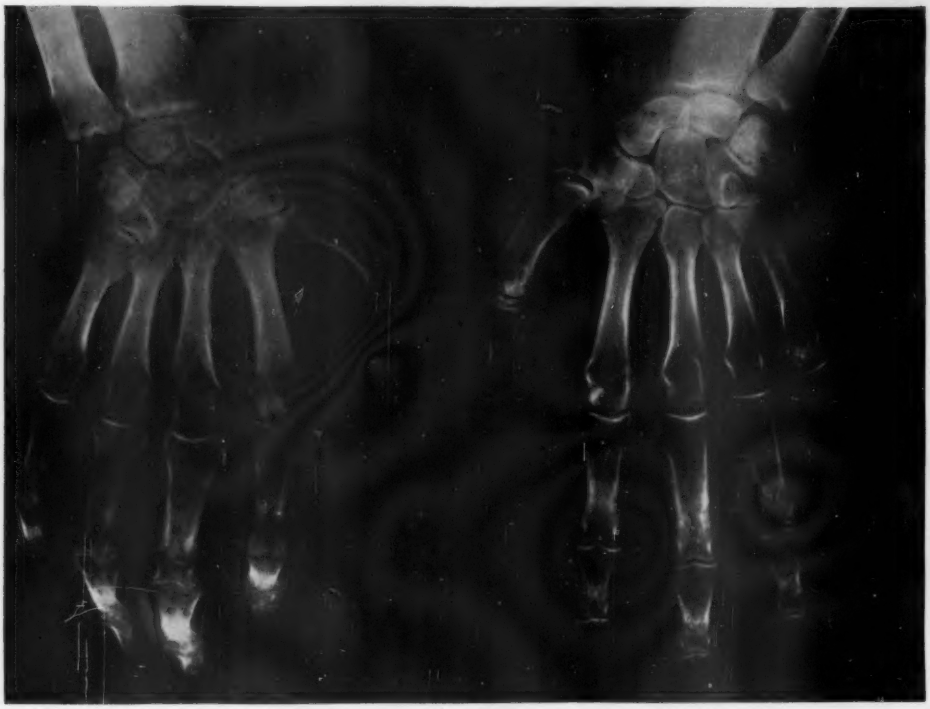

FIG. 11

Case 2—Well marked Sudeck's changes in the wrist and hand. There is typical spotty atrophy involving the carpus and the ends of the metacarpals and phalanges. The increase in soft-tissue shadow of the palm can be seen. The view of the last two phalanges of each finger is distorted because it is impossible to straighten the fingers. There is an old destructive lesion of the second metacarpo-phalangeal joint.

whereas Dupuytren's contracture is at least six times more common in men, may be explained by the higher incidence of wrist fractures in women. None of our patients was a known epileptic and none had knuckle pads (Skoog 1948).

SUMMARY AND CONCLUSIONS

1. The incidence of Sudeck's syndrome in the hand is about once in 2,000 accidents of all kinds.
2. The onset of signs and symptoms occurs some weeks after injury and the typical radiographic changes (spotty atrophy) appear six to eight weeks later.
3. Conservative treatment by heat, elevation and graded function is effective if initiated within six weeks of the onset of symptoms.
4. There is some close relationship between Sudeck's atrophy and thickening of the palmar fascia.

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HEREDITARY FORMS OF RICKETS AND OSTEOMALACIA

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It is becoming clear that the forms of rickets and osteomalacia now met with in this country are almost invariably due to primary metabolic abnormalities rather than to environmental factors. This does not imply an absolute increase in this metabolic form of disease, but only its uncovering by the virtual disappearance of primary nutritional deficiency of vitamin D.

One may perhaps think of classical rickets as it was seen fifty years ago as being largely due to environmental causes, or nurture, and rickets as we see it today as due largely to genetical causes, or nature. The former has been effectively eliminated by an alteration in the diet; the latter seems to develop independently of the living conditions in which the individual is brought up and appears to depend on some inborn biochemical peculiarity in the make-up of the individual.

At the present time, in this and in most other civilised countries, nature has become the more conspicuous factor and therefore apparently the more important since many of the problems of nurture have been largely solved. This demands a considerable reorientation of our approach to our patients' diseases. It is the reason why increasing attention is now being paid to medical genetics and why there is increasing acceptance of the fact that some individuals are constitutionally different from others and develop differently and have different requirements for health even in the same environment.

Nevertheless a sharp distinction between these two factors so far as concerns their contributions to disease processes has never been and can never be complete. In the early days of the study of the pathogenesis of classical rickets hereditary factors had often been suspected (Siegert 1903, Pfaundler 1911, Ziesch 1925) and twin studies, notably by Lehmann (1936), confirmed that such factors were of significance. It is likely that different children require somewhat different amounts of vitamin D to maintain optimal growth and that such variations are, no doubt, at least in part due to differences in their genetical constitutions. When there is an adequate diet such differences in vitamin requirement will be unimportant because the vitamin will be present in excess of requirement for practically all children. However, when the vitamin content of the diet is at a low or borderline level, rickets is likely to become a relatively common disorder and the genetical differences between individuals will become more significant in deciding why one child and not another develops the disease. Thus, while the frequency of classical rickets in a population or stratum of a population may depend largely on the environmental conditions, it could be mainly genetical factors which decide which particular children within the population are going to get the disease. In the past, however, looking at the disease as a whole, these genetical factors never appeared very conspicuous. In the last thirty years, however, vitamin D intake is so much higher than it used to be that the only cases which still get a disease similar to classical rickets are ones in which heredity appears all important.

It is interesting here to note the parallel studies recently done on another action of vitamin D, namely the toxic action. It now appears that some people are far more sensitive than others to vitamin D toxicity, being severely affected by ordinary small anti-rachitic

doses. This serious disease has only been uncovered recently by the very factor (increased vitamin D intake) which has caused classical rickets to disappear. The sensitivity to vitamin D may occur in infants for an unknown reason (Thatcher 1936; Bonham Carter, Dent, Fowler and Harper 1955), and is the cause of some cases, at least, of the syndrome described by Lightwood (1952) as " idiopathic hypercalcaemia of infancy with failure to thrive." The sensitivity almost certainly occurs also in some adult cases of sarcoidosis (Anderson, Dent, Harper and Philpot 1954; Henneman, Carroll and Dempsey 1954). So far, hereditary factors have not yet been implicated here.

In this paper we are considering some of the forms of rickets and osteomalacia which still occur in spite of this high average intake of vitamin D in our population. Most of these cases have been described under the name of " vitamin D resistant rickets " in the past for the excellent reason that they are very similar indeed to classical rickets, except that the curative dose of vitamin D is many thousand times greater than is the dose which is anti-rachitic for the vast majority of people.

In an earlier paper to this Journal (Dent 1952) these forms of rickets were described and classified in six main groups according to the degree of abnormality of renal tubule function which was present. This was, and still is, a convenient classification for clinical purposes because it draws attention to a disorder of function which can be readily detected and measured, which is possibly responsible for the bone disease, and which has to be corrected, partly at least, by therapy. The same basis of classification has been independently used by Fanconi (1951) and by Jackson and Linder (1953). In our earlier paper various objections were made to the use of the term " vitamin D resistant rickets," although it was conceded that the classification on the basis of tubule function was also inadequate since it gave no account of other metabolic abnormalities, many of which were similar to those occurring in vitamin D deficiency and which were located elsewhere than in the renal tubule. These and other objections to both forms of classification still hold. The most valid objection to the term " vitamin D resistant rickets " is that it can be equally well used to describe the similar vitamin D resistance occurring in other diseases such as steatorrhoea and renal-glomerular failure. This objection we now think has become stronger, for recent work in our department suggests that the mechanism of the vitamin resistance may be similar in all these cases. On the other hand we must now withdraw one of our earlier objections, namely that " vitamin D resistance " could not explain the frequent association of the bone disease with other congenital tubule defects " which have nothing to do with vitamin D " (Dent 1952). We are now beginning to believe that some of these tubule defects, like the phosphate defect, have a great deal to do with vitamin D action because they are sometimes reversed with the massive vitamin D therapy used to cure the bone disease.

We will also discuss here our present attempts to distinguish the various clinical types of rickets and osteomalacia of metabolic origin and our preliminary views as to the importance of heredity in their pathogenesis. These studies are beginning to show that there are very many more distinct forms of these diseases than we had previously suspected, and that the problem of classification and nomenclature gets more and more difficult. We propose, therefore, to describe the diseases as such, under rather ponderous names, without committing ourselves to a means of classification. In most of the diseases described our family studies have shown the importance of hereditary factors. However, we include for the sake of completeness some other clinically similar variants of these diseases in which we have so far been unable to prove the influence of heredity. These latter diseases are very rare and we may well uncover hereditary factors when we have been able to study more cases. Cases in which the bone disease appears to be the result of steatorrhoea of any kind or of primary renal-glomerular failure will not be included since the bone disease (and therefore the orthopaedic problem) in such cases constitutes only a minor part of the clinical syndrome and since the influence of heredity in such cases is not yet sufficiently obvious to be of clinical or theoretical value.

Likewise we will make no mention of the forms of rickets and osteomalacia due apparently to renal-tubular acidosis with nephrocalcinosis (Types V and VI of Dent 1952), since these likewise are apparently not hereditary. However, we must mention here in passing the fascinating hereditary form of nephrocalcinosis described by Schreiner, Smith and Kyle (1953). This is probably a completely new syndrome and is not primarily a bone disease. Unfortunately we have no experience of this condition.

We rather fear that a review of this kind of a group of rare medical and orthopaedic conditions will be of only limited interest. We have been encouraged to complete this work by the classic study of Fairbank (1951). His very detailed *Atlas of General Affections of the Skeleton*, written mainly from a surgical point of view, has proved of extraordinary value to workers in other branches of medicine. We, who are medically and biochemically biased, are now attempting in our small way to continue in the tradition set by this great man using the excuse that this important occasion gives us, and hoping that it too will be of value outside our own very limited field of activity.

DESCRIPTIONS OF THE VARIOUS FORMS OF RICKETS AND OSTEOMALACIA IN WHICH HEREDITARY FACTORS ARE PROVED OR STRONGLY SUSPECTED

In the following descriptions of the various syndromes we have included data mainly obtained from our own patients but sometimes also from patients kindly referred to us by others but remaining under their care. Unless otherwise stated, however, we have been responsible ourselves for the genetic survey. It may be possible later to publish data in more detail on the individual syndromes concerned. We can only hope here to prepare the way by briefly describing the syndromes and by giving representative genetical data. It will be noted that the syndromes divide themselves into two main groups, the first being patients without renal failure in the ordinary sense of the word but all having nevertheless a renal-tubule defect of phosphate reabsorption. These correspond to the Types I–III of Dent (1952). This defect, as well as some of the other associated biochemical and clinical defects, many of which may also mimic vitamin D deficiency, can be corrected partly at least by treatment with large doses of vitamin D. In the second group (Type IV of Dent 1952) the same defect of renal tubule reabsorption of phosphate with or without other findings reminiscent of vitamin D deficiency is also present. There is usually a marked renal glycosuria and aminoaciduria. However, these patients have other defects also of renal function, such as proteinuria, inability to secrete a dilute or concentrated urine, a variable, usually slight, degree of nitrogen retention, and so on.

Most of these syndromes have been described by others under various names in the literature, the first group mainly as " vitamin D resistant rickets " and the second as " renal rickets " or " Fanconi syndrome." One of the hereditary diseases we are mentioning here (hypophosphatasia) is not strictly speaking a true rickets according to our usual histological and biochemical definitions. It is included here, however, because the clinical appearances, and to some extent the radiological ones, can cause some confusion with rickets.

FLORID RICKETS PRESENTING IN INFANCY WHICH CLOSELY MIMICS DIETARY DEFICIENCY OF VITAMIN D BUT WHICH REQUIRES ENORMOUS DOSES TO PRODUCE HEALING

This is the most common form of rickets nowadays in this country and comprises most of the cases of renal-tubular rickets described, as of Type I (Dent 1952),* or by others as refractory or vitamin D resistant rickets. We have not much data from early life but the little we have suggests that these patients are quite normal clinically and radiologically at birth, and grow and develop normally for the first year of their lives. After this, however, usually

* It is greatly regretted that our theory of renal-tubular defect in this syndrome and of the mechanism of the curative action of vitamin D was put forward in our original publication without knowing and therefore acknowledging that the identical point of view had been previously expressed by Robertson, Harris and McCune (1942).

before the second year, the child is noticed to be slow in attaining its milestones, and may then show unambiguous rachitic deformities. This is not very conspicuous at first, however, and many are not brought to see the doctor until three or four years of age, by which time dwarfism, lateness in walking, abnormalities of gait and bow legs may have become quite obvious, especially to somebody who sees them for the first time. By then the radiographs will show a florid rickets. Biochemical investigations show a normal plasma calcium, low inorganic phosphate, raised phosphatase and normal bicarbonate. The usual renal function studies give normal results and in particular the urine contains no excessive quantities of aminoacids. As likely as not the patient will have received ample doses of vitamin D from early childhood (1,000–2,000 units a day) and probably will have received larger doses (10,000–20,000 units a day) from other doctors before being referred to us. We suspect that a slight response at first on the somewhat higher range of dosage may sometimes be observed but that this is at best only very temporary. Such a patient has no other biochemical abnormalities that are easy to determine. More detailed investigations, however, also show faulty calcium absorption from the gut and low urinary calcium excretion.

This condition, once developed and left to itself, is constant and incurable. On account of the normal general health of the patients, however, they survive to adult life, when untreated, with varying degrees of deformity, which has usually necessitated osteotomy (Fig. 1). By about fifteen years of age they will be greatly dwarfed (Pedersen and McCarroll 1951), with limbs very small in comparison to the length of the trunk, which is almost normal. They are likely to get aches and pains in their bones during childhood, and to be rather easily tired. When growth has ceased, however, the subjective clinical signs become less conspicuous. Biochemically they maintain the same findings described above, except that the alkaline phosphatase level may return to normal. From the age of about twenty to forty the patient may be symptom-free. Some time after this, however, the pains and radiological signs of osteomalacia are likely to appear and this may be severe enough to cause complete crippling.

It is important perhaps to point out a few differences that there are between this condition and classical rickets due to lack of vitamin D. The main difference, apart, of course, from the dietary history and natural course of the disease, is that in this form of rickets tetany never occurs. The plasma calcium level may be slightly low, somewhere between 9·0–10·0, but never falls to tetany levels (less than 7·5) as it so frequently does on and off for some obscure reason in a case of classical rickets of long enough duration. Another difference concerns the muscular weakness and hypotonia, which is so typical of classical rickets and is not very conspicuous in this form of refractory rickets. Indeed, some of these patients, even when untreated, are exceedingly muscular and vigorous, especially when they reach adolescence. Another difference is that cases of classical rickets may have a mild aminoaciduria (Jonxis 1955), unlike the majority of cases of this syndrome.

The treatment of this condition requires the administration of very large doses of vitamin D, something of the order of 5–10 milligrams a day of oral calciferol (1 milligram=40,000 units) being required to cure the radiological signs and to maintain them cured.

FIG. 1

Photograph of an adult patient of fifty-four with the severe form of rickets and osteomalacia. Most of his deformities occurred in childhood and were several times corrected temporarily by osteotomy. He was symptom-free from the age of twenty. Recently backache and other pains returned and he thought the deformities were worsening. Until this picture was taken he had had no medical treatment. He is the patient I 1 in family tree R 1 of Figure 4.

Under this treatment the metabolic disorders may be entirely corrected (Fig. 2) and much more satisfactory growth and avoidance of deformities obtained, although even then patients tend to end up somewhat dwarfed (Fig. 3). There is great danger of vitamin D intoxication because the toxic dose appears to be only a little more than the dose required to cure the rickets. This is a curious situation, since most normal patients can only be intoxicated by

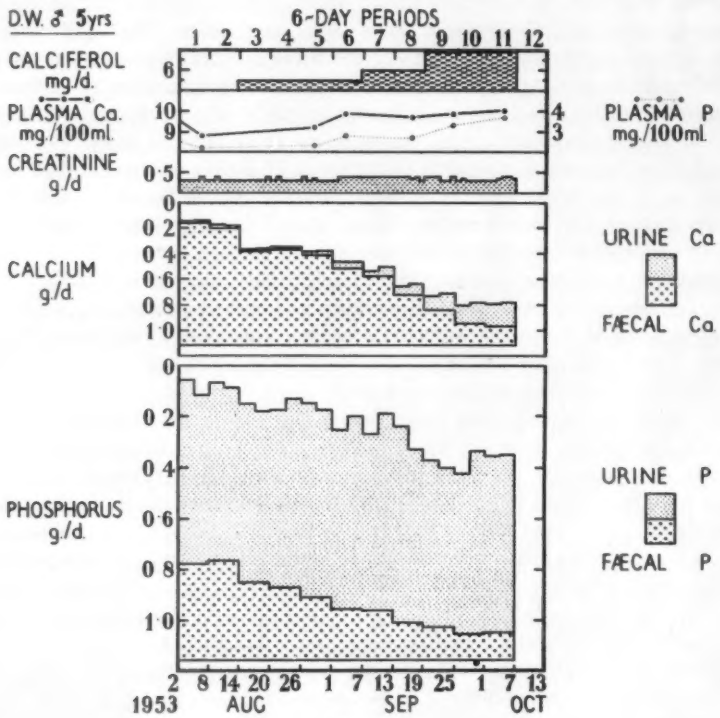

FIG. 2

Metabolic data in a boy of five years with the severe and commonest form of hereditary rickets. The results of giving calciferol are shown. The intake of calcium and phosphorus is measured downward from the zero line: the output of faecal and urinary calcium is measured upwards, hence the clear space below the zero line represents the positive balance or retention of calcium or phosphorus. Before treatment he has a high faecal calcium, low urine calcium and is in smaller positive balance than he should be for his age. These changes are identical with those found in classical rickets. On giving very large doses of calciferol faecal calcium falls considerably. There is a slight rise in urine calcium and he goes into strongly positive calcium balance. Phosphorus balance changes are parallel, but are only half as big as the calcium. This is because calcium phosphate is being laid down as bone in the usual Ca:P ratio of approximately 2:1 respectively. Plasma levels also become nearly normal during the period of treatment. These changes are also identical with those occurring after treatment of classical rickets.

similar large doses of vitamin D, while their anti-rachitic requirement of calciferol is very small indeed. The " resistance " to vitamin D in this disease concerns, therefore, its anti-rachitic action not its toxic action. This is one of the reasons why we do not like the term "raised resistance to vitamin D" (RRD) introduced by McCance (1947) to describe this and related syndromes.

The treatment of an adult case who presents with symptoms is exactly the same as for the infant, the dose of vitamin D being about the same, and also its hazards. It is easier to define the biochemical criteria for the treatment of a fully grown adult. Our working rule is that we should try and raise the plasma phosphorus to more than 2·5 milligrams/100 millilitres

with a dose of calciferol which does not raise the plasma calcium to more than 11·5 milligrams/100 millilitres, nor raise the urine calcium output to more than 400 milligrams/24 hours. When toxicity is developed we prefer to stop treatment altogether until this has been fully recovered from and then to start again with the calciferol.

This treatment of children and adults is only moderately satisfactory and we are further investigating ways of improving it in order to obtain a more normal clinical response with less dangers of toxicity. We hope to publish some improvements in treatment not yet fully confirmed in a further paper. We still cannot make up our minds what to do about a symptom-free patient in the middle age group. It will require very painstaking studies to determine their true status and how far energetic treatment then may avoid later troubles. Usually we do not treat them unless suspicious symptoms appear.

FIG. 3

A girl of twenty-four with the severest and commonest form of hereditary rickets but who otherwise enjoys perfect health. She is the daughter of the patient shown in Figure 1, and is Case II 1, in family tree R 1 of Figure 4. She was well treated with fairly large doses of vitamin D in the early days of this treatment (she was case J. L. of MacKay & May 1945) and has avoided most of the deformities that her father has had and has required no osteotomy. However, it will be seen that her limbs are relatively short in proportion to her trunk and her stature is small, being about the same as that of her younger normal brother (II 3) of ten years, seen by her side.

The condition is frequently found in more than one member of the same family, often in different generations. Probably two out of every three affected individuals are derived from parents one of whom is similarly affected. In the remainder of cases both parents appear to be quite normal. A proportion of the children of affected patients also tends to have the same condition. Typical pedigrees are shown in Figure 4.

This kind of familial configuration is consistent with the hypothesis that the disorder is due to a rare abnormal " dominant " gene and the affected individuals are heterozygous for this. Probably the gene is incompletely manifesting: that is to say a proportion of those carrying it does not for some reason show the condition. This could explain why some cases are derived from parents both of whom are apparently quite normal, and why on our present rather small figures it appears that rather less than one in two of the offspring of affected patients are also affected. More convincing proof of such incomplete manifestation would be provided by families in which the condition appeared to skip a generation, but so far we have not encountered such a situation ourselves, nor in the literature.

Another possible explanation of the occurrence of apparently sporadic cases, or of families in which a series of affected individuals in more than one generation appears to have originated from a pair of healthy parents (*e.g.*, Family R.8) is the occurrence of fresh mutations. While it is likely that such fresh mutations do occur, and presumably balance the loss of the gene from the population due to the somewhat diminished effective fertility of patients with the disease, it is rather difficult to decide at the moment what proportion of cases do in fact arise in this way.

Considerably more data will be required before the full details of the genetics, the rate of manifestation, and the proportion of cases due to fresh mutation can be established.

The fact that an appreciable proportion of the offspring of an affected patient (probably at least 30 per cent) is liable in due course to develop the disease is of considerable therapeutic importance. If all such children were kept under observation from birth it should be possible to identify those who are going to be affected at a very early stage of the development of the condition. This would enable active therapy to be begun early enough to prevent any deformity developing and perhaps to result in quite normal growth.

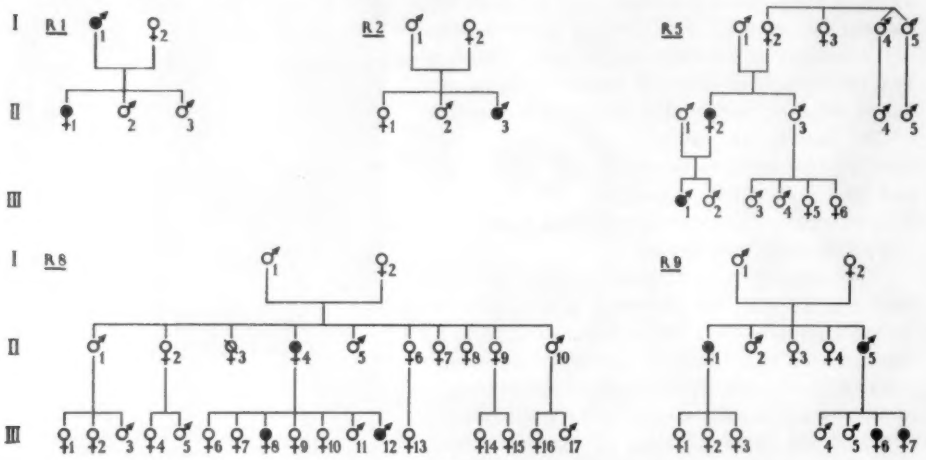

FIG. 4

Representative family trees (R 1, R 2, R 5, R 8, R 9) of some of our cases of florid rickets presenting in infancy. In this and subsequent pedigrees affected members are shown in black; a line drawn obliquely through the circle means died in infancy.

If it is true that the clinical disorder is not manifest in a proportion of individuals carrying the abnormal gene, a number of problems of considerable physiological interest are raised. It might be that careful study would reveal slight deviations from the normal in such individuals and enable them to be recognised and further investigated with a view to discovering why no gross physiological disturbances have occurred. So far our preliminary attempts to identify such individuals on the basis of their heights and bodily proportions, or on the levels of their plasma inorganic phosphate, have not been successful. It seems possible that if such deviations do occur they will only be detected by rather more refined procedures. Studies of renal-tubular function with respect to phosphate where both parents are apparently normal would be of obvious interest.

MILDER RICKETS PRESENTING IN CHILDHOOD WHICH CLOSELY MIMICS DIETARY DEFICIENCY OF VITAMIN D AND REQUIRES MODERATELY LARGE DOSES TO PRODUCE HEALING

It is interesting that we have seen a very few examples of a syndrome which is almost identical with the one described above except that we suspect that the manifestations are rather milder and that the curative dose of vitamin D is much less. Other authors, such as Pedersen and McCarroll (1951), in their excellent review, have also suggested that there are varying degrees of severity in resistant rickets. In this milder form it may be that the parents may have noticed nothing wrong with the child till four or five years of age, or even later. In some cases the rachitic deformities have been corrected by osteotomy and no further complaints have been made apart from lack of stature until adult life, when at the age of thirty or over signs and symptoms of osteomalacia appear. The biochemical findings are exactly the same in blood and urine as in the previous syndrome, except that we suspect that

the plasma phosphorus may be a little less low and more variable. The main difference, however, is in the response to treatment, for good clinical and radiological cure can be obtained with 0·5–1·0 milligram a day of oral calciferol, whereas, as already mentioned, 5–10 milligrams are required for the more severe form of this disease.

We have only been able to study two families with patients of this sort. So far as our data go it appears that the condition runs true to type in the particular families, so that all the affected members in a family appear to require similar treatment. In one family a woman aged fifty-four, her son aged twenty-eight and the son's daughter aged five, all showed the condition. In the other a mother and her daughter were affected, whereas a son was normal. It is likely therefore that the condition is inherited in much the same way as the more severe type, and that the affected individuals are heterozygous for a rare abnormal gene. Whether the disease assumes a less severe form in these cases because a different gene is concerned, or whether it is the same gene and other factors lead to a modification of its expression, remains to be determined.

SEVERE OSTEOMALACIA PRESENTING IN EARLY ADOLESCENCE OR ADULT LIFE AND VERY CLOSELY MIMICKING DEFICIENCY OF VITAMIN D

This is an exceedingly rare syndrome with only a few other descriptions like it in the literature, as for instance the case described by McCance (1947). We have studied carefully four cases of this disease: two were girls, presenting in the late 'teens; and two were men, presenting in their twenties. In the case of the youngest member the first sign was that from the age of twelve years she ceased to grow. A waddling gait developed two years later. However, unambiguous osteomalacic aches and pains were not noted until she was sixteen. At the age of twenty she had gross radiological changes of osteomalacia with softening of bones and Looser zones. The other girl presented more acutely at the age of nineteen with aches and pains, muscular weakness and a waddling gait, and showed well calcified bones radiologically but with multiple Looser zones. The two men presented rather similarly but showed marked loss of height as possibly the first symptom. All four patients had straight limbs, and otherwise showed no stigmata of previous rickets.

It is rather characteristic of these cases that very marked muscular weakness is present, and indeed in one of our cases it was so gross that the patient lay immobile in bed unable to raise his head off his pillow and scarcely able to move his arms and legs. The first diagnosis made in his case was of progressive muscular atrophy, and we have heard of other similar cases which had been for some time confused with myopathies and neurological diseases. Apart from all this, however, the patients enjoy excellent general health. Biochemically they are very similar to the two previous types, already described, of refractory rickets with a plasma calcium usually between 9 and 10 milligrams per cent, phosphorus of 1–2 and a raised alkaline phosphatase. Like the other cases too, these patients do not get tetany.

There is a further curious finding which we are in the course of investigating and which was present in all four of our cases, namely a moderately lowered plasma bicarbonate level. This is a most important fact to consider in the diagnosis since this finding is usually taken to indicate the presence of an acidosis, and an acidosis with osteomalacia or rickets at once makes us think of other syndromes, such as are usually described under the names of renal-tubular acidosis. The main basis of treatment of this latter is with alkalis rather than with large doses of vitamin D; so it would be unfortunate to confuse the two. The chief clinical difference is that in true renal-tubular acidosis there is usually other evidence of renal damage, such as slightly raised blood urea, inability to acidify the urine, proteinuria, nocturia, defective concentration and dilution of urine. All these are absent in the syndrome under discussion, where the only detectable renal defect is diminished renal-tubular reabsorption of phosphate. Another biochemical difference is that the twenty-four-hour urinary calcium excretion is lower than normal in our syndrome and higher than normal in renal-tubular acidosis.

This is a very serious condition and three of the four patients were almost completely crippled by the time they were referred to us. The treatment is with very large doses of vitamin D, starting with, say, 5 milligrams a day of calciferol and raising the dose as necessary, judging mainly by the plasma phosphorus levels, and with the same working rule mentioned above that the plasma phosphorus should be raised to over 2·5 milligrams/100 millilitres, the calcium not over 11·5, and the urine calcium not more than 400 milligrams/24 hours. This latter stipulation is particularly difficult to uphold in this syndrome and the patients are therefore in considerable danger of renal stone formation. One of our patients had to receive 20 milligrams a day of calciferol for a long time, and we are continually faced with the problem that the curative dose for the rickets is often a dose which produces hypercalcaemia with its attendant complications. Indeed, one of our cases, treated and intoxicated already elsewhere, showed paradoxically enough signs of both ectopic calcification and of osteomalacia at the same time. Treatment of these patients has not been too satisfactory in the past but we are very hopeful that we are greatly improving it at the moment by using different forms of vitamin D. This work is not ready for publication. It is interesting that the muscular weakness which plays so large a part in the clinical syndrome disappears on treatment with vitamin D. The plasma bicarbonate usually rises a little on the vitamin D, sometimes to normal or low normal levels.

An interesting biochemical finding has been that all four of our patients had a high excretion of one aminoacid—glycine—in their urine. We allude to this matter again later in this paper.

In none of the families of these patients have we observed any other relatives to be affected nor was there any parental consanguinity. However, with such small experience of the disease we cannot exclude a hereditary factor and prefer in any case to mention it here among the proved hereditary diseases because of its very close clinical and biochemical relationship. It is of great interest that McCance's original patient (1947) has now apparently made a spontaneous recovery after having initially required large doses of vitamin D. We understand that she has now maintained herself well for some years without treatment, without symptoms and with normal plasma, calcium, phosphorus and phosphatase levels (McCance, private communication).

OTHER ASSOCIATED FINDINGS IN RICKETS AND OSTEOMALACIA OF ABOVE TYPES

While nearly all the cases we have seen of non-acidotic rickets or osteomalacia can be included in one or other of the above three categories, it is only too painfully obvious that the matter is indeed far more complicated than this and that it will only be a matter of time before we have to extend further the descriptions of distinct syndromes. As an indication of the way this may go we will now mention some further findings in some of these diseases.

Osteomalacia has now been described as a rare concomitant of neurofibromatosis (Hernberg and Edgren 1949, Dent 1952, Swann 1954; Saville, Nassim, Stevenson, Mulligan and Carey 1955). This association appears rather too frequent to be fortuitous and suggests some common causal relationship. The osteomalacia presents in adult life and closely resembles the severe adult presenting form described above. It is not correlated with any particular clinical form of neurofibromatosis. Neurofibromatosis is well known to be inherited. It is determined by a rare abnormal gene for which the patients are heterozygous. It is markedly variable in its manifestations. Whether or not the syndrome of neurofibromatosis with osteomalacia represents a rare variant determined by the same gene, or whether we are concerned with an entirely new situation, cannot be decided at the moment because of the paucity of family material, and because of our ignorance of the underlying biochemical disturbance which leads to neurofibromatosis. We are not aware of a case of neurofibromatosis associated with proven refractory rickets in childhood.

FIG. 5

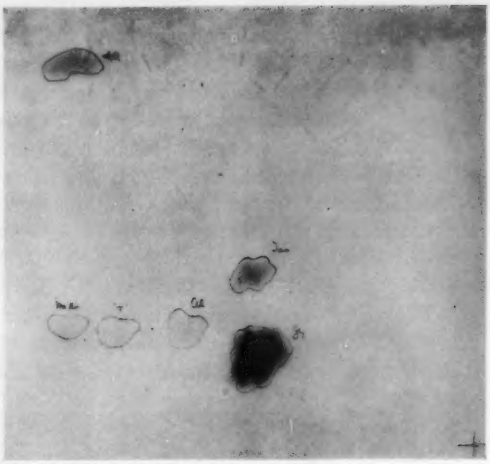

FIG. 6

FIG. 7

FIG. 8

Paper chromatograms of urine showing main aminoacids present. The measured amount of urine has been placed on the cross at the lower right-hand corner of the square of filter paper, and phenol and then collidine have been soaked across the paper in the directions from right to left, and upwards, respectively (Dent 1948). The aminoacids are then revealed as purple spots on the paper, each one in its appointed place. They may be identified by reference to a published " Map of the spots " (Dent 1951). Figure 5—Normal urine. This specimen shows an aminoacid excretion at the upper limit of normal. On such a specimen the quite normal " glycine pattern "—so called because the strongest spot is due to glycine—can be easily seen and compared with the pathological urine patterns. Figure 6—Urine from a patient with severe osteomalacia presenting in early adolescence. This shows the " super-glycine pattern " mentioned in the text characterised by disproportionate excretion of one aminoacid only—namely glycine—which is seen as the largest spot on the paper and which is much stronger relative to the other spots than it is in Figure 5. Figure 7—Urine from a case of Lignac-Fanconi disease or cystinosis. The chromatogram shows a large number of strong spots widely distributed over the paper which indicate the generalised aminoaciduria characteristically found in these patients. This was the patient III 4, shown on family tree F 3 of Figure 15. Other data from this patient are in Figures 10 to 12. Urines from the adult form of this disease show an identical aminoacid pattern on the chromatogram. Figure 8—Urine from a case of hypophosphatasia. The only conspicuous abnormality here is in the lowest spot shown on the chromatogram. This spot is due to phosphoethanolamine, a substance never found in normal urine by our methods. The remaining spots appear at the upper limit of normal strength owing to the fact that more urine than usual was taken for analysis on this chromatogram.

Craniostenosis has now been several times described together with our first form above of florid rickets presenting in infancy (Imerslund 1951, Coleman and Foote 1954). It is of interest that Case 7 of Bickel, Smallwood, Smellie and Hickmans' (1952) series of Lignac-Fanconi syndromes also had craniostenosis. This child showed many features not typical at all of the Lignac-Fanconi syndrome, and indeed he resembled much more closely our first form of florid rickets. It is curious that such premature closure of sutures should ever, even if rarely, occur in a disease characterised by poor calcification and retarded bone age.

Albinism occurred in a case of florid rickets presenting in infancy described by Winberg, Bergstrand, Engfeldt and Zetterström (1954). The same authors described another case with a similar form of rickets, this time, however, associated with alopecia totalis. One of our own cases (unpublished) has also had this latter complication.

The remaining associated findings all concern renal-tubule function abnormalities. They are of special interest in providing evidence relating to theories of etiology, whether for instance primary tubule dysfunction is the cause of the disease, or whether it is due to a more generalised disorder of vitamin D metabolism. It seems that the phosphate reabsorption defect in the renal tubule can be equally well explained in either of these two ways. The association of these other tubule abnormalities does make it almost impossible at the moment to explain it on the basis of the third possible theory, namely that it is due to secondary hyperparathyroidism. This latter is probably the most popular theory at the moment for accounting for the phosphate defect, but in our opinion it does not explain the known facts.

Renal glycosuria is now a well established feature of some cases of non-acidotic rickets and osteomalacia (Lièvre, Bloch-Michel, Sassier and Solignac 1948; Dent 1952). Its theoretical interest led one of us to classify these cases under a different name from the others (Type II of Dent 1952). Unfortunately we have not yet had such a case under our own clinical control. All these cases, and indeed the other types of hereditary rickets and osteomalacia also, require accurate measurement of the renal tubule reabsorption maximum (T_m) for glucose. Until this is done, both before and after treatment with vitamin D, we will remain in some doubt as to its significance.

Renal aminoaciduria has been known for some time to be a feature of the acidotic forms of rickets and osteomalacia to be described later. Less commonly it may occur without acidosis (Type III of Dent 1952) and in the only case followed up for some time we have noted that the aminoaciduria disappeared on treatment with vitamin D. A case of this syndrome has been well described by Kyle, Meroney and Freeman (1954). In these cases the aminoaciduria is generalised and concerns most of the commonly occurring aminoacids. Only recently, and at the instigation in the first instance of Dr P. Astrup, did we notice that a further abnormality could also occur which involved one aminoacid only—glycine. These patients excrete large quantities of glycine without any increase whatever in the other aminoacids. The pattern of aminoacid excretion as seen on paper chromatograms is rather characteristic (Fig. 6). It has been found in all four of our cases of severe late-presenting osteomalacia (above) but also in some few cases of the florid rickets presenting in infancy. We are unable to interpret these findings at the moment. Our theoretical interest in such aminoacidurias must now be reassessed in the light of the recent work of Jonxis (1955) on classical rickets. He has reported a renal aminoaciduria as occurring in vitamin D deficiency, which largely disappears on treatment with vitamin D. The pattern of aminoacid excretion in classical rickets is quite different from that which may be found in any of our hereditary forms of rickets and osteomalacia. We must take note, nevertheless, of the new action of vitamin D that has been revealed by this work.

THE SO-CALLED " ORGANIC ACIDURIA " SYNDROME OF LOWE, TERREY AND MACLACHLAN

A most interesting disorder was described by Lowe *et al.* (1952). Its main features were " organic aciduria," decreased renal ammonia production, congenital glaucoma, and mental

retardation. However, of the three patients described two had rickets which was readily cured with large doses of vitamin D and alkalis, and all three had rarefaction of their bones. The syndrome was complex, for there were also behaviour abnormalities, aminoaciduria, proteinuria, intermittent glycosuria and a chronic acidosis. The urinary findings closely resembled those of the Fanconi syndrome, although clinically the two diseases are quite different.

We have been fortunate enough to diagnose a further example of this syndrome in a patient who was admitted to University College Hospital under Dr B. Schlesinger. This child was twelve years old and presented with a florid rickets with dwarfism and gross deformities and he also had most of the other signs described by Lowe *et al.* It is hoped that this case will be published fully later. We mention it here because of its exceptional interest in that an older brother of the child had obviously had the same syndrome but had unfortunately died some years ago during orthopaedic treatment (Fig. 9). There was no evidence in the case reports of Lowe *et al.* that the syndrome was hereditary.

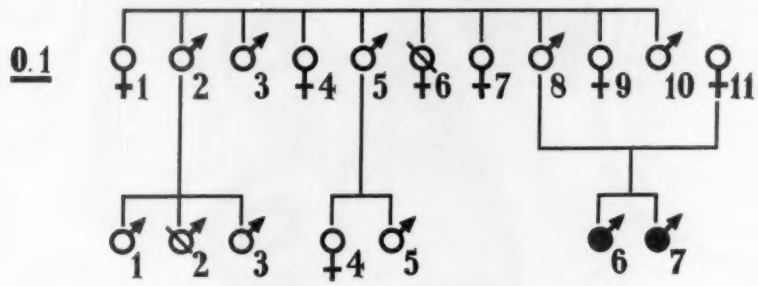

FIG. 9
Family tree (0.1) of a case of the so-called " organic-aciduria " syndrome of Lowe, Terrey and MacLachlan (1952).

We have not completed our biochemical studies, but so far as they go they are similar to those of Lowe *et al.*, the only conspicuous difference being that we have not confirmed the excretion of increased amounts of organic acid as distinct from aminoacid.

LIGNAC-FANCONI DISEASE OR CYSTINOSIS (DE TONI-FANCONI SYNDROME, FANCONI SYNDROME, Etc.)

The clinical picture of this most interesting disease has been dealt with rather thoroughly (Bickel, Smallwood, Smellie and Hickmans 1952) so it is not necessary to deal with this in any detail here. The children present usually at the age of about two years with failure to thrive, backward development and rickety deformities. The marked differences between this disease and those which have so far preceded it in this paper is that the children are iller and have many of the signs of chronic renal failure, namely feeding difficulties, thirst, lassitude and a great susceptibility to infections or to febrile reactions of unknown origin. They often get tetany. For this reason the patient is much more likely to be seen first by the paediatrician rather than, as in the other diseases reviewed here, by an orthopaedic surgeon. A preliminary examination would show a slightly raised blood urea with normal or low plasma calcium, phosphorus and bicarbonate levels. Phosphatase is usually raised. The urine is constantly dilute and contains from traces to large amounts of protein. It is clear that at first the vague diagnosis of " renal rickets " is likely to be applied to these cases together with the hopeless prognosis that this description usually implies. It should have been noted, however, that the plasma phosphorus level is lower than would be expected for this degree of renal failure, and further examination of the urine will show the presence of glucose and

of large quantities of aminoacids (Figs. 5 to 8). It is important, too, and usually easy, to examine the cornea with a small beam of light and hand lens for the presence of cystine crystals, which appear in the form of small gritty particles. Under slit lamp examination and with a magnification of thirty to fifty times, the individual crystals can be seen as tiny needles. This latter finding is pathognomic of the disease. The radiographs, if taken after one year of age, show florid rickets without any suggestion of secondary hyperparathyroidism. The disease represents one of the best examples of severe renal-tubule damage with only relatively small damage to the renal glomerulus. The disease is therefore characterised by excessive loss of substances from the body rather than by retention of excretory products and it is this that makes it easier to treat than other forms of renal failure, for the treatment merely comprises

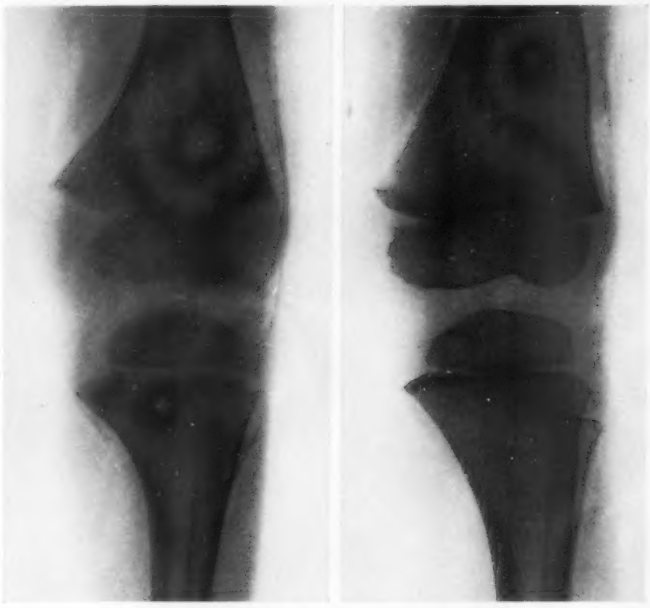

FIG. 10 FIG. 11

Radiographs of knees of one of our patients with Lignac-Fanconi disease before and after seven months' treatment. The times correspond to the two arrows shown in Figure 12. It is seen that marked growth has occurred of normally trabeculated bone. The original coarsely trabeculated rachitic bone with its lines of arrested growth can be seen in Figure 11 farther along the shafts beyond the recently laid down normal bone. There is also a slight bulge there corresponding to the original splayed-out metaphyses. These radiographs are identical in appearance with those that may be obtained from cases of classical vitamin D deficiency rickets before and after treatment.

giving by mouth quantities of such substances as are being lost in the urine in sufficient amounts to cause a metabolic derangement, together with a sufficient dose of vitamin D to correct some of the other metabolic defects (Dent 1952, Bickel and Smellie 1952). Under this treatment the rickets can be cured (Figs. 10 and 11) and the child's symptoms almost completely alleviated. In the past all these children died young from severe infections or electrolyte crises. We do not yet have sufficient follow-up data to indicate what the long-term prognosis is but they can certainly be made very much better for many years. It appears at the moment that after three or four years growth slows down (Fig. 12) and renal-glomerular damage increases, so that unless improved forms of treatment are forthcoming they will probably die in uraemia in childhood. It is interesting that Bickel and Thursby-Pelham (1954) have shown

that some at least of the signs of tubule damage are not present at birth. Although most of the evidence for tubule damage when it is found is of a biochemical nature, it is to be noted that Clay, Darmady and Hawkins (1953) have put this on a morphological basis by their microdissection studies of the nephrons from these cases. They have shown that there is, as might be expected from the biochemistry, a gross abnormality of the proximal tubule (Figs. 13 and 14). This is shorter than normal and is joined to the glomerulus by a narrow swan-like neck.

The genetics of the disease has already been discussed in a preliminary fashion by Bickel and Harris (1952). Subsequent work has supported their preliminary conclusions that the disease was due to an autosomal recessive gene. Typical family trees are shown in Figure 15.

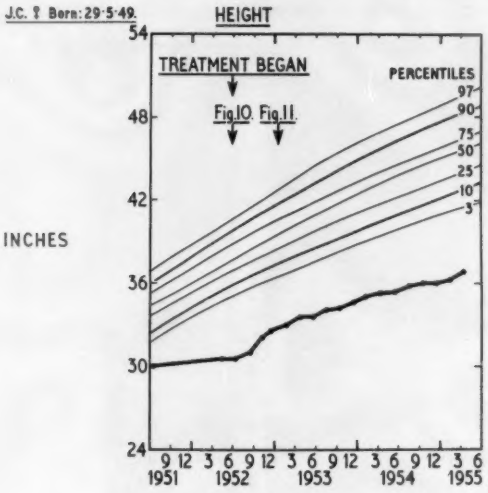

FIG. 12

Growth curve from a case of Lignac-Fanconi disease. It can be observed that she is very dwarfed as against the normal percentiles for her age and that she hardly grew at all when she was receiving no treatment during the nine months before being sent to us. When treatment began a spurt of growth occurred which is actually faster than normal for that age, as shown by the slope of the line being steeper than normal. After a year, however, growth began to slow down. This is our usual predicament.

Note the occurrence of the disease in only one sibship in each family and the first cousin parents in F 5 and F 7. The disease is remarkably homogenous, especially among affected members who are siblings. According to Bickel and Harris (1952) all of their fourteen cases of the disease had cystinosis—that is, there was microscopical evidence of cystine deposition in the body. In our view one occasionally sees cases, clinically and biochemically identical with theirs, in whom it is not possible to prove the presence of cystine storage. Post-mortem material from such a case is necessary for detailed chemical analysis as a negative microscopic finding is difficult to interpret. So far, post-mortem material has been only from cases in which clear cystinosis had been diagnosed in life. Analysis of the spleens of these patients shows that 3 per cent or so of the wet weight is pure cystine. This is up to a thousand times more than the normal quantity (Cusworth, Dent and Fowler, unpublished data).

MILD CHRONIC RICKETS AND OSTEOMALACIA WITH MANY SIGNS OF RENAL-TUBULE DYSFUNCTION

Recently we have been able to recognise that there is a group of cases that are different from those with Lignac-Fanconi disease, although resembling it closely in many respects. In this group the rickets is usually much less likely to be a presenting sign. More often it is the symptoms of renal damage which are first complained of. There may be feeding difficulties, vomiting, " acidosis " dating from early life, and usually there is a very gross thirst and polyuria. The rickets, if it does present at all, is likely to do so later in childhood or adolescence than in the Lignac-Fanconi syndrome, but clearly some mild degree of rickets is present from an early age since dwarfism is a constant feature. We have studied carefully only four of these cases and have good records of a fifth from another hospital. In all cases but one the renal damage was not detected in childhood and the rickety deformities, if any, had been

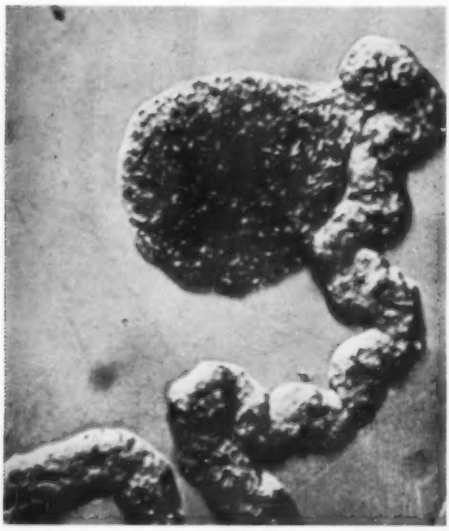

FIG. 13

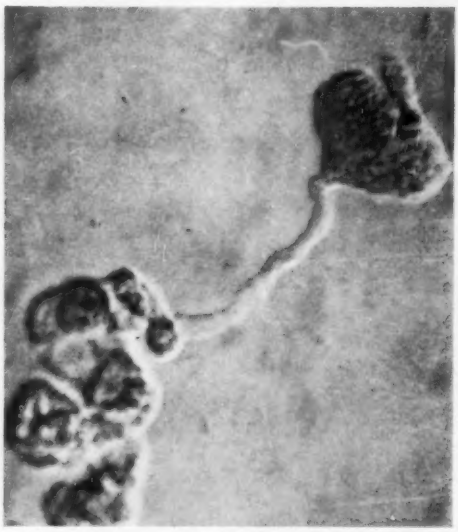

FIG. 14

Photomicrographs of dissected nephrons taken from Figures 2 and 5 of Clay, Darmady and Hawkins (1953). The glomerulus and the first part of the proximal tubule is shown in each case. A normal nephron (×160) is shown in Figure 13; a nephron (×95) from a case of Lignac-Fanconi disease (Case III 2, of family F 7 of Figure 15) is shown in Figure 14. Note that in this Figure the first part of the proximal tubule adjacent to the glomerulus is grossly abnormal.

corrected by osteotomy without further note of the disorders of renal function present. The two cases that have reached adult life have then presented at the age of thirty and forty years with the signs and symptoms of osteomalacia, but even then their shortness of stature indicates the long duration of some bone disease and careful history-taking has always elicited an almost life-long disease. Investigation shows biochemical findings identical with those in the Lignac-Fanconi disease except that these cases do not have cystinosis and that their urine calcium excretion is higher and not, as in the other syndrome, lower than normal. The treatment is based on the same general principles as in the other forms of renal-tubule disease: one replaces electrolytes lost in the urine, such as sodium bicarbonate, and gives sufficient vitamin D to correct the other abnormalities, especially the low plasma phosphorus level. It is of special interest that two patients have died with cirrhosis of the liver and a third patient we have under observation has a slightly enlarged hard liver with apparently normal function. We originally described cases such as this under the name " Fanconi syndrome "

and wondered if the cirrhosis of the liver was due to the chronic loss of aminoacids in the urine (Stowers and Dent 1947). Our present view is that cirrhosis of the liver does not occur in the other syndromes mentioned here, many of which also have gross aminoaciduria. The cirrhosis is not, therefore, very likely to be due to the aminoaciduria, However, it may still be that the duration of the aminoaciduria is longer in this syndrome than in the others, and that therefore the aminoacid depletion is also greater.

As yet we have no evidence that the condition is hereditary but more cases must be studied before this can be considered further. The disease is certainly present soon after birth and does not progress much subsequently. Hence, if the renal damage is acquired rather than hereditary then it must be a very curious form of renal damage. In one of our cases, a short-statured boy who presented at the age of thirteen with mild rickets and knock-knee, we have some evidence for the presence of a urethral valve with some obstruction to urinary flow. It is unfortunate that no nephron dissection studies have yet been reported from cases such as this.

"FANCONI SYNDROME" PRESENTING FIRST IN MIDDLE AGE

Of all the problems of nomenclature in the various forms of rickets it is in this syndrome that things are most confused. However, we and others have used the term "Fanconi syndrome" for an adult syndrome, which is quite extraordinarily similar to that occurring in children and more correctly given this name. As there is no better name for it we prefer to continue calling it thus for the time being.

The disease presents around the age of forty as a typical osteomalacia of the more acute type (Dent and Hodson 1954) in which the bones become tender, show Looser zones on radiography, but otherwise appear of fairly normal structure and density. The patient may have noted some loss of height and after some time a moderate degree of muscular weakness also develops. He does not get tetany, and renal symptoms, while always present, may be inconspicuous. Biochemically the plasma levels are typical of those of an acidotic osteomalacia with normal or slightly lowered calcium, low phosphorus, low bicarbonate and raised phosphatase. The plasma potassium is usually a little lowered, as occurs in other forms of renal-tubular acidosis, and one of our patients (the same one as first described in detail by Milne, Stanbury and Thomson 1952) also had quite typical attacks of low potassium paralysis. The urine, however, is the most interesting feature. It contains large quantities of aminoacids and glucose just as in the children's form of this syndrome, and there is a defect of acidification whereby a load of ammonium chloride by mouth does not produce an acid urine, at least until the plasma bicarbonate level has been very greatly lowered. There is also protein in the urine and other tubule function defects such as inability to concentrate or dilute adequately. These adult cases of the Fanconi syndrome do not have cystinosis as detected microscopically or by chemical analysis of organs. Another marked biochemical difference between the syndrome and the Lignac-Fanconi disease is in the urine calcium output. This tends to be lower than normal in the children with Lignac-Fanconi disease but is usually greatly raised in the adult syndrome. Despite these differences it must be pointed out that the apparently highly characteristic morphological defect of the proximal tubule of the nephron is the same in both child and adult forms of the syndrome (Clay, Darmady and Hawkins 1953).

This disease is difficult to diagnose clinically, although it is exceedingly easy to diagnose biochemically once the possibility has been considered. Clinically the patient presents with symptoms that are not at all specific. He may be feeling some backache or complain of rheumatic pains in his feet or hips. As his bones show no gross rarefaction or deformities radiographically, and as the Looser zones may not appear early in the disease and even when present may be very difficult to find, it is easy to see how patients may be undiagnosed until they are seriously ill. By our genetic studies we have been able to detect cases of this syndrome before the occurrence of clinical signs of disease. These patients have later presented with

quite severe bone pains which showed nothing on radiographic examination but which completely disappeared on giving the specific treatment. Such cases are quite undiagnosable by ordinary means. We refer to this matter again in the discussion.

Treatment has been very satisfactory. It comprises giving doses of sodium or potassium bicarbonate and of vitamin D sufficient to correct the metabolic abnormalities, using the

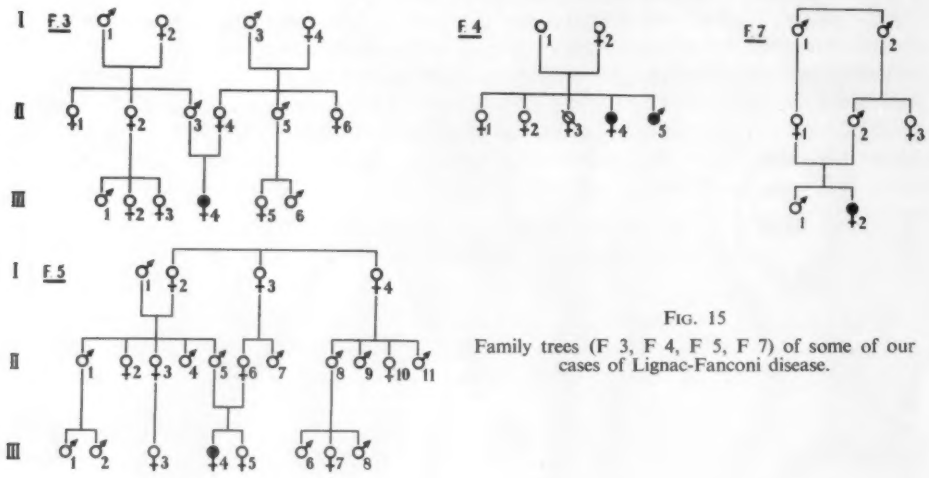

FIG. 15

Family trees (F 3, F 4, F 5, F 7) of some of our cases of Lignac-Fanconi disease.

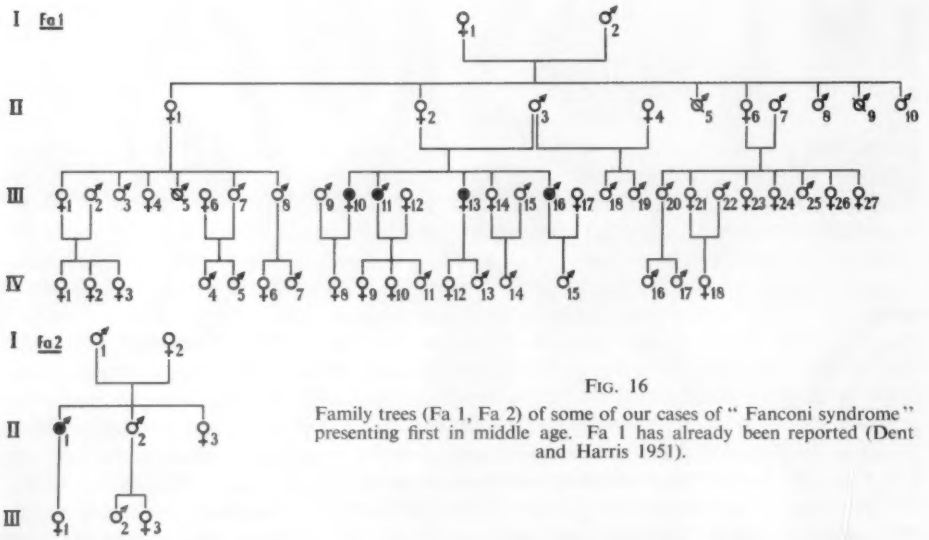

FIG. 16

Family trees (Fa 1, Fa 2) of some of our cases of " Fanconi syndrome " presenting first in middle age. Fa 1 has already been reported (Dent and Harris 1951).

same criteria as in the treatment of other forms of acidotic osteomalacia. The high urine calcium which these patients have has to be rather carefully watched in case it rises further with the vitamin D. It is a curious fact, however, that these patients to our knowledge have never formed stones nor developed nephrocalcinosis, in spite of the fact that this is usual with patients who have other forms of renal-tubular acidosis and in whom the same features predisposing to stone formation are present, namely neutral urine with a high calcium content.

We wonder whether the aminoaciduria in this syndrome exerts a protective action against stone formation. This is possible on theoretical grounds.

Two of the families in which this condition has occurred are shown in Figure 16. The simplest hypothesis is that the condition is inherited as a Mendelian recessive character. Its relation to the childhood form of the syndrome remains a difficult problem. In so far as the two conditions run true to type in individual families it seems that different abnormal genes must be involved. It would be of interest to know whether these genes were at different chromosomal loci, or were alleles. If they were found to be alleles this would probably mean that the series of underlying biochemical disturbances leading to the clinical syndrome would be qualitatively very similar though different in degree.

HYPOPHOSPHATASIA

This most interesting hereditary biochemical disorder of bone was first described as such by Rathbun (1948) in an infant in whom there was defective calcification of bone with softening of the skull and deformities of the limbs superficially suggesting rickets. Biochemically the blood showed raised calcium, phosphorus and non-protein nitrogen levels, with low, almost undetectable alkaline phosphatase levels. The infant died and the bones were found to show osteoid hyperplasia with disorganised calcification as in severe rickets. However, the suggestion was made that in this condition the osteoblasts were unable to produce alkaline phosphatase and that this was the primary defect and the cause of the bone disease. Since then a few more cases have been described (Sobel, Clark, Fox and Robinow 1953, Schlesinger, Luder and Bodian 1955), and the original findings of Rathbun confirmed and extended. It seems likely that the condition is hereditary, because relatives of the affected patient often show chronically low alkaline phosphatase levels while being apparently clinically normal. The genetic studies of this condition are not, however, yet suitable for publication. The affected children's general health is comparatively good and it is beginning to appear that they make a spontaneous recovery after a few years. Many cases of this syndrome may have been already described under other names—for instance Case 2 described by Anspach and Clifton (1939), in which the suggestion was made that the child had hyperparathyroidism on account of its raised plasma calcium level. Cases which may well have been of hypophosphatasia have also been described as having unusual forms of renal rickets (Chown 1936, Schneider and Corcoran 1950). We wonder too whether some of the cases described as atypical chondrodystrophies may not have been cases of hypophosphatasia. It is rather fun to try to rediagnose cases that have been fully enough described in the past. Of especial interest to us was the adult patient with fragile bones and a very low alkaline phosphatase described by Macey (1940). We think it is quite likely that this case was an adult survivor of the children's disease because he had had the "rickets" in childhood and had got much better in later childhood. In the children's disease it appears that the diagnosis can be reliably made by paper chromatographic analysis of the urine for aminoacids (Cusworth and Dent 1955). This does not reveal an aminoaciduria but a gross abnormality of rather unusual kind, namely a very large excretion of phosphoethanolamine (McCance, Morrison and Dent 1955, Fraser, Yendt and Christie 1955), a substance which is not detectable in normal urine (Fig. 8). Dr Henneman (Boston) has kindly sent us recently a urine specimen from an adult with fragile bones who resembles very closely clinically the patient described by Macey. We were intrigued to note that his urine showed exactly the same abnormality as the children with hypophosphatasia. It appears that it may be possible to make a little more sense soon out of an obscure corner in orthopaedics —namely the problem of fragile bones and of some obscure bone dystrophies.

OTHER RARE CAUSES OF OSTEOMALACIA

Osteomalacia may occasionally arise as a consequence of a true metabolic disorder occurring in a much more complex clinical syndrome. In such situations the reason for the infrequent association of osteomalacia with the more obvious primary disorder present may

at first sight appear rather obscure. It is possible that something of this sort happens in neurofibromatosis, as has been mentioned earlier. Probably a clearer example is the case of Wilson's disease.

This neurological disorder is well known to be hereditary, and it is now clear that the affected patients are homozygous for a rare abnormal " recessive " gene (Bearn 1953). The clinical manifestations are variable in onset, rarely beginning in childhood, more commonly in adolescence, and occasionally delayed still further. It is now thought possible that the underlying biochemical disturbance is a failure to synthesise in normal amounts the copper-containing plasma protein ceruloplasmin (Scheinberg and Gitlin 1952). This failure for some reason leads to an excessive absorption of copper from the gut (Cartwright *et al.* 1954) and to its consequent slow deposition in the tissues throughout life. Copper is deposited most extensively in the brain and liver, and the characteristic lesions that develop can be attributed to the toxic effects of the large amounts of copper which slowly accumulate in these sites. The variable onset of the clinical condition is presumably due to variable rates of copper absorption from the gut and possibly to variations in the copper content of the diet. The condition illustrates very clearly one aspect of the nature-nurture problem. The individual has an inborn incapacity to cope adequately with one specific aspect of his environment, namely the copper content of the diet.

Among the diverse features of this disease there has recently been described renal-tubule dysfunction. This can probably be attributed like the other lesions to the toxic effects of excessive copper deposition. Usually aminoacid reabsorption is most markedly disturbed (Cooper, Eckhardt, Faloon and Davidson 1950), but dysfunction in glucose and uric acid reabsorption has also been encountered in a proportion of cases. It is probable that phosphate reabsorption is also occasionally involved because some cases of Wilson's disease have been described (Warnock 1952, Cooper *et al.* 1950) with radiological and biochemical signs of osteomalacia. The tubular damage is clearly very variable both in onset and extent, and this is no doubt because it is a rather remote reflection of the fundamental disorder of copper metabolism, attributable to the presence of a particular abnormal gene in double dose. The occasional occurrence of osteomalacia in Wilson's disease can thus be understood as a rather distant consequence of an underlying genetical abnormality.

Similar renal-tubule damage and osteomalacia has also occurred in what is usually accepted as an acquired disease. This is the most interesting case of multiple myelomatosis described by Engle and Wallis (1954), in whom, after some years, typical renal-tubule abnormalities developed—renal glycosuria, aminoaciduria and acidosis—with subsequent development of the typical syndrome of adult osteomalacia with multiple Looser zones.

Hence it appears clear that the same bone disease can be caused by the same renal-tubule disorder, even when of widely different etiology.

DISCUSSION

We have described the various forms of rickets and osteomalacia which we have been able to study clinically and genetically and in most of which we consider hereditary factors to be mainly responsible for the disease. The great majority of the cases we have seen belong to one or other of the forms we have described. Perhaps we should summarise here those features of more immediate clinical value which may have arisen from this study.

In the first place, we hope that we have made clear that there is often excellent treatment for hereditary diseases. There is still some misunderstanding of this matter: for instance, even certain British authors have stated in a paper describing a case of the Fanconi syndrome: " The condition being a congenital and often hereditary defect of the renal tubules, treatment is unfortunately of little avail." It is our view that the hereditary or acquired nature of a disease has no bearing whatever as to whether or not we can, or at least can hope to, treat it adequately. With this more optimistic approach to the study of hereditary diseases, we can

now make a further obvious point that, in adisease in which progressive bone or other damage occurs, the sooner the diagnosis is made the better. Here the hereditary diseases enjoy a great advantage over acquired ones, since it is only in them that the onset of a disease can sometimes be to some extent predicted. This point is illustrated in our case history (described briefly by Dent and Harris 1951) and subsequent follow-up (unpublished) of a family (Fa 1 of Fig. 16) in which a case of the adult Fanconi syndrome had been diagnosed. The patient (III 13) was completely crippled when we first saw her and gave a seven-year history of increasing pain, weakness and general misery. It took over six months' medical treatment before she could be returned to her normal activity. Our family study showed that three of her four siblings had the biochemical features of her disease while having no complaints whatever. These three have been kept under observation and have all subsequently developed clinical signs of the disease, which we have now been able to treat very effectively within a very short time of its appearance. In the youngest member of this sibship the clinical signs only developed recently, that is, five years after we had first detected his biochemical abnormality on the original family survey. It is clear, therefore, that whenever one of the diseases described in this paper is diagnosed in a patient, it is our responsibility to examine as many as possible of the remaining members of the family, however symptom-free they may appear at the time. It is here that a knowledge of the mode of inheritance is of great advantage.

In the case of recessive disorders one in four of the brothers and sisters may be expected on the average to develop the condition. The risks so far as the children of a patient are concerned are on the other hand very low, unless the patient happens to have married a first cousin. In the case of dominant or partially dominant conditions there is a risk approaching one in two, according to the rate of manifestation, that the brothers and sisters of an affected individual will develop the disorder, and a similar risk that the children of such patients will also be affected. The development of appropriate screening tests to detect such predisposed individuals is one of the most important tasks of research in this field. In the Fanconi syndromes routine examination of the urinary aminoacids has proved the most useful to date. In the other syndromes the investigation of young children with respect to the phosphate, calcium and phosphatase levels in the plasma, and appropriate radiography of the bones, would seem at present to be the most useful way of identifying the potentially affected individuals at the earliest possible time. We cannot stress too much the fact that true hereditary diseases need not be congenital. They may be manifested at any time from foetal life until well on in later life.

With regard to the help which one can get from genetic studies in making a more accurate diagnosis of patients with rickets or osteomalacia, there are several points to be made. The most important is, of course, that the clinical diagnosis must be one which is consistent with the known facts of its inheritance. The occurrence of an entirely different mode of inheritance of a condition seemingly identical clinically with a form previously recognised would imply some difference in causal relationships. One would be suspicious, for instance, if the diagnosis of Fanconi syndrome had been made in both a parent and child of the same family in the absence of consanguineous marriage. The next point concerns the rather remarkable homogeneity of each disease when it does recur more than once in a family. This not only helps in diagnosis but it also means that the results of extensive investigation and treatment of one of the patients can be used directly to benefit the others. This is particularly noticeable with regard to such points as the dose of vitamin D that is necessary to cure the disease. A further way of saying the same thing is that if in a family with a diagnosed case of bone disease another patient with bone disease presents himself, these diseases are almost certain to be the same. We had an interesting example of this from a mistake brought to our notice on one occasion. This concerned a woman who had radiological signs of pathological fractures and abnormal biochemical findings. She was diagnosed as having a parathyroid tumour and was twice, without success, explored surgically, once in the neck and once in the chest, for the tumour.

It was not until after this that it was noticed that her daughter had been under treatment for some years at a children's hospital for one of the forms of metabolic rickets. It then at once became clear that the mother really had the same disease. This was confirmed and the mother is now doing well on the same medical treatment as her daughter.

Many further problems remain in studies of this kind which we should like to mention in the hope that others will be better able to deal with them. One of these concerns the problem of orthopaedic correction of deformed bones in patients whom we know we can treat adequately by medical means. It is most important that attempts should be made to correct deformities of the vertebrae, for instance, when the disease is first diagnosed, so that when the bones have been strengthened the patient ends up in more normal shape. It may be feasible too to straighten bent malacic long bones and deformed pelves before treatment hardens them.

Another unsolved problem concerns a more purely genetic matter, that of detecting heterozygotes in the diseases in which we know the mode of inheritance to be recessive. By a comparison of the disordered metabolism of heterozygotes and homozygotes we may be able to determine the effects of an abnormal gene in single and double dose. Furthermore the development of methods for detecting such individuals would enable us to predict more accurately the chances of further children being born with the disease. We are often asked about the future children of apparently normal relatives of patients, for instance, with the Fanconi syndrome. Their chance of being themselves heterozygous for the abnormal gene is very high, but of course they will not bear abnormal children unless they should happen to marry another person who is also heterozygous for that gene. Clearly they are likely to do this if they marry into their own family or into any other family which has an affected member, and we can advise them of the probable risk should they wish to do this. Should they marry into an apparently normal family, however, we have no way of telling what may happen, although the risk fortunately is then always rather small. It is quite likely that very sensitive tests for the abnormalities present in the homozygous patients could be developed which would be capable of detecting the heterozygotes but, except possibly in hypophosphatasia, this most desirable objective has not yet been achieved in any of the recessive diseases discussed here.

SUMMARY

1. We have described here various forms of rickets and osteomalacia that we have studied ourselves and have come to recognise as comprising definite syndromes. We have included only diseases in which hereditary factors have been proved or might be suspected to play a part.

2. There are more of these syndromes than previously defined.

3. All these hereditary diseases can be treated, usually very satisfactorily, using the same general principles and with only very few simple medicines such as vitamin D and sodium bicarbonate. Careful control and long follow-up, however, may be required to achieve best results and to avoid accidents.

4. A study of the genetics of the diseases is a help in diagnosis and treatment. Broadly speaking the group of diseases which mimic vitamin D deficiency more or less closely tend to be inherited in dominant fashion. The groups with more severe degree of renal-tubule abnormality are inherited as recessives.

5. When more than one member of a family is affected it has been found that in each family each syndrome runs true to type.

We sincerely thank the clinicians, too numerous to mention by name, who, by referring cases to us, have enabled these investigations to be carried out; and who have also helped us in very many other ways. We wish to thank also Miss Barbara Warland for her assistance in the family case work.

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THE VALUE OF KNOWING THE DIRECTION AND NATURE OF THE FORCE CAUSING A FRACTURE

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From St Thomas's Hospital

Medical knowledge, expanding from day to day, is becoming so large that we should hesitate to add to the things that have to be learnt, and we may well ask ourselves whether anything is to be gained from knowing the direction and nature of the force causing a fracture. Yet this knowledge is of value, in the first place because we can deduce from it the degree of damage to the soft parts, and in the second place because we can infer from it a logical method of reduction.

The damage sustained by the soft parts often controls treatment. For example, in a crush injury, such as is commonly associated with a motor-cycle accident, the soft tissues suffer much and the skin may be cut or scalped; in a tapping type of injury with little momentum, such as happens when a man is kicked by a horse, the skin is often cut but the soft parts are not otherwise damaged. The difference in the degree of soft-part injury affects immediate treatment. In a crush injury tissues may be devitalised and a wound-toilet operation is often desirable on that account; in a tapping injury there is not any devitalised tissue and a wound-toilet operation is therefore unnecessary. Again, soft parts are not damaged in a spiral fracture and the bone may safely be plated on the day of the accident, whereas in a crush fracture it is advisable to postpone plating for a fortnight.

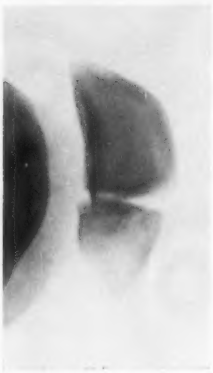

FIG. 1
Fracture of patella, caused by indirect injury.

In the case of a fractured patella the nature of the injuring force decides between two dissimilar treatments. The indirect fracture of the patella occurs when an elderly patient loses his balance. Instinctively he contracts his quadriceps and the extensor apparatus of the knee gives way. The result is a clean transverse fracture of the patella, with rupture of the quadriceps expansion on either side (Fig. 1). From the point of view of treatment the injury ranks as a torn tendon, the ends of which have to be approximated and sewn together. The direct fracture of the patella occurs when a person of any age strikes the ground with his knee cap. The patella breaks into several fragments (Fig. 2). There is no muscle damage and the several fragments are held together by the surrounding intact quadriceps, as Furlong says, " like toffee in its paper " (personal communication). Neither muscle nor bone demands treatment and the patient is handled as though he had only sprained his knee. As soon as he can lift his leg off the couch with the knee straight he is encouraged to get up without a splint (Fig. 3).

FIG. 2
Fracture of patella, caused by direct injury.

THE EVIDENCE OF THE RADIOGRAPH

To those who can interpret the signs, the radiographs of a fracture nearly always reveal the direction of the force that broke the bone. A penetrating high-velocity missile has a shattering effect; it behaves as though it exploded after entering the bone, and small fragments are displaced in all directions (Fig. 4). This kind of fracture is rarely seen except during a war. In civilian life the possible forces are longitudinal compression, angulation, shear, transverse compression and rotation.

Longitudinal compression occurs for example when a person falls from a height and lands on his feet. It causes either several longitudinal linear cracks without displacement, or a T-fracture into a joint. The patient shown in Figure 6 fell from a height; he has a fracture of the calcaneum on one side and vertical cracks in the tibia on the other. The patient shown in Figure 5 fell on the point of her flexed elbow.

FIG. 3
Range of knee movement four weeks after the fracture shown in Figure 2.

FIG. 4
Gunshot fracture.

The term impaction is much misused. Complete impaction—one end of the bone burying itself in the other—as a result of a compression force is rare, but if the bone is angulated there may be impaction on one side, as in the abduction type of transcervical fracture of the neck of femur (Fig. 7).

An *angulation force* causes a transverse fracture of the shaft of a bone, often with a separate triangular fragment, the presence of which indicates that the bone was bent until it broke (Fig. 8); but it also shows the direction in which it was bent, for the triangular fragment is always on the inside of the bend. In a child angulation often causes a greenstick fracture.

A force that strikes the shaft of a bone at right angles over a limited area and with dying momentum may be called a *tapping force*. It causes a clean transverse fracture, often without displacement or at the most with a mild amount of shift. The man shown in Figure 9 was kicked by a horse. With continued momentum in a transverse direction the force may be described as a *shearing force* (Fig. 19).

FIG. 5
Longitudinal compression: T-shaped fracture into elbow.

FIG. 6

FIG. 7

Figure 6—Longitudinal compression: linear fracture of tibia (R), fracture of calcaneum (L).
Figure 7—Angulation force: impacted fracture of the neck of the femur.

Transverse compression occurs when a limb is crushed. The nature of the injuring force is self-evident when a bus wheel runs over a leg or a man is pinned under a collapsed building; but crushing is also present in many motor car accidents. The crushing element is important, for the soft tissues are damaged as well as the bones, and the degree of damage to the soft parts often controls the immediate treatment. It was possible to deduce from Figure 10 that the tissues had been crushed; plating was therefore postponed for fourteen days, the patient meanwhile being made comfortable by skeletal traction. Several days after the accident the damage to the soft parts became self-evident (Fig. 11).

FIG. 8

FIG. 9

Figure 8—Angulation force: transverse fractures of the tibia and fibula, at the same level. Figure 9—Tapping force: transverse linear fracture of the tibia.

Rotation causes a spiral fracture. The fracture is restricted to the diaphysis but extends over a considerable stretch of the shaft. The common fracture of the shaft of the femur in children is produced by rotation. In the adult, rotation damages the leg below the knee more often than the thigh; the tibia is broken in the lower third and the fibula in the upper third, and both fractures have the characteristics of a spiral fracture (Fig. 12). An oblique fracture is a variant of the transverse fracture, and is caused either by angulation or by shear. In the oblique fracture it is possible by taking several radiographs to find one that shows a gap throughout the whole extent of the fracture. In a spiral fracture it is not possible to get a radiograph that shows a complete gap unless the fragments are widely separated. Figure 13 at first sight looks like a spiral fracture; it is in fact an oblique transverse fracture.

FIG. 10
Crushing force: oblique fractures of tibia and fibula at the same level.

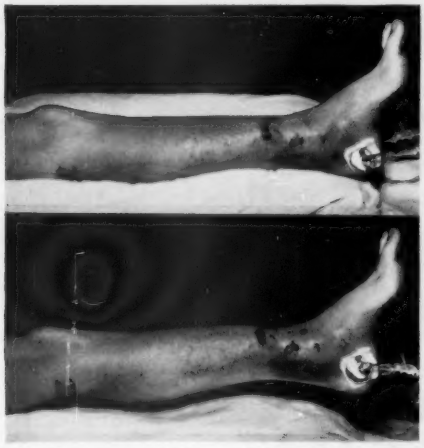

FIG. 11
Seven days after the fracture shown in Figure 10. Patient lifting his leg off the bed. Note injury to the soft parts.

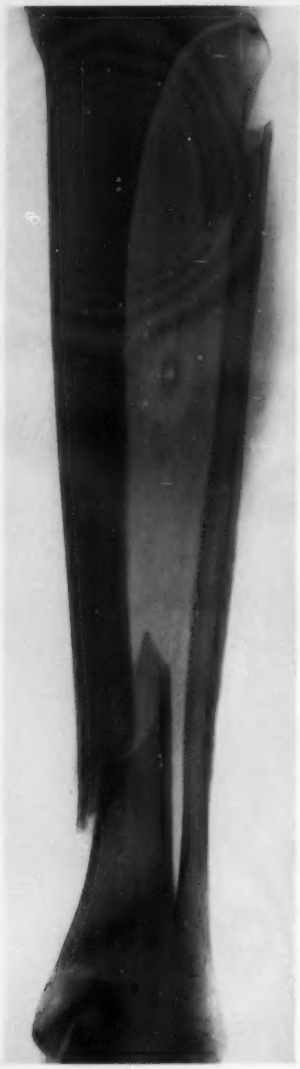

FIG. 12
Twisting force: spiral fractures of the tibia and fibula at different levels.

OTHER EVIDENCE

In addition to radiography there are other pointers to the nature of the fracturing force.

The history is important but it may be equivocal. A person falling from a height and landing on his feet has obviously been exposed to a longitudinal compression. When, however, a child falls on to its outstretched hand and sustains fractures of the shafts of the radius and ulna, a rotational force in addition is involved, for as the child strikes the ground the trunk twists on the fixed hand.

The nature of the skin damage is informative. After a head injury a graze on the face implies that the patient came to rest by sliding rather than by butting something head on, and damage to the brain is not likely to be serious. A wound on the forehead makes one suspect a hyperextension injury to the neck. And experience tells us that if a patient has hurt his foot treading off the pavement he has in all probability sustained a lateral-rotation Pott's fracture.

FIG. 13
Oblique fracture mimicking a spiral fracture.

In many fractures a knowledge of the direction of the fracturing force points out an obvious and easy method of reduction. For example, in a Colles's fracture the lower fragment has been tilted backwards by an angulation force and, at the same time, it and the hand have been supinated on the ulna. To correct the displacement an opposite force is applied: the doctor's two thumbs are placed on the back over the lower fragment of the radius, and the lower fragment is tilted forwards and pronated (Fig. 14). As another example Mervyn Evans (1951) has pointed out that fractures of the forearm caused by a fall on the hand can be reduced by supinating the forearm.

THE FORCES CAUSING POTT'S FRACTURES

A Pott's fracture best illustrates the usefulness of knowing how a bone has been broken. The fracture Pott described is different from the fracture he sustained, and the exact nature of both is uncertain; but it is convenient, clinically, to use the term Pott's fracture to cover any fracture of the leg bones involving the ankle joint.

It is customary to describe degrees of Pott's fracture. This is to be deprecated on two accounts. First, there is not any point in distinguishing a fracture as first, second or third degree unless the method of treatment is different; and second, the terminology gives one no clue to the direction of the fracturing force and therefore no clue to the force required to reduce the displacement.

Subdividing Pott's fracture according to the direction of the fracturing force we have the following varieties: lateral-rotation, abduction, adduction, shear and vertical. And in nearly every case radiography enables the doctor to guess correctly which fracture he has to deal with.

Lateral-rotation causes a fracture through the lateral malleolus. The fracture is often invisible in the antero-posterior radiograph; in the lateral it appears as a line, more vertical

FIG. 14
Method of reducing Colles's fracture.

FIG. 15
Lateral-rotation Pott's fracture.

than horizontal, which begins below at the level of the mortise and runs upwards and backwards (Fig. 15). The medial malleolus may or may not be broken; if it is, the antero-posterior radiograph shows a transverse fracture below the level of the horizontal part of the ankle mortise. The situation of this fracture is important because the portion of the medial malleolus left attached to the shaft acts as a buffer against which the talus can be reduced. The lateral malleolar fragment is displaced backwards; when the displacement is great the posterior third of the articular surface also splits off from the tibia. This fracture is seen as a vertical line in the lateral view. In most lateral-rotation Pott's fractures the displacement is slight; only when the displacement is great does it reveal clearly that rotation has been the essential fracturing force (Fig. 16).

A lateral-rotation fracture is reduced by rotating the foot inwards. Exact replacement of the bones is not necessary provided that the talus has been restored to its original bed in the ankle mortise. As in all Pott's fractures, subluxation of the ankle, not the displacement of the bones, is the important fact and treatment is centred on restoring the talus to its proper position. The amount of displacement of the talus varies from a slight lateral shift or a slight lateral tilt to almost a dislocation. But whatever the amount of the displacement, the method of reduction is the same. There is no point therefore in subdividing a Pott's fracture into degrees, as is often done.

The *abduction fracture* is not as common as the lateral-rotation fracture, but is more important because it is much more likely to leave the ankle crippled. In the antero-posterior radiograph there is a transverse fracture of the fibula about two inches up (Fig. 17). Often a small intermediate fragment indicates by its triangular shape that the break was caused by angulation. The medial malleolus may escape or it may break at the same level as in the lateral-rotation fracture.

Displacement in an abduction fracture is corrected by adducting the foot. To make the reduction complete considerable force is required. One need not be afraid of over-reduction because the small fragment of the medial malleolus that remains attached to the shaft acts as a buttress to prevent the talus from overshooting the mortise.

The abduction fracture, being horizontal, is prone to angulate when the patient starts to bear weight. For this reason an abduction Pott's fracture needs protection while it is proceeding from union to consolidation, whereas in most other Pott's fractures splintage can safely be discarded as soon as the bones have joined.

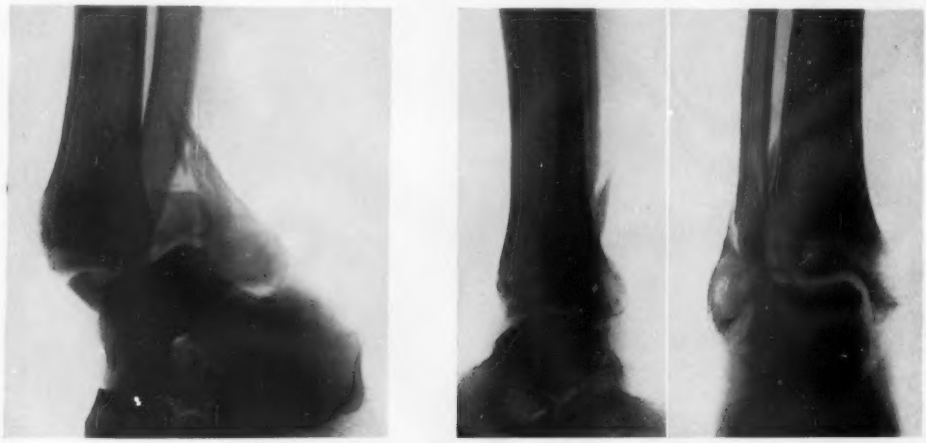

FIG. 16
Lateral-rotation Pott's fracture before and after reduction.

An *adduction* force on the foot causes as a rule a sprain of the subtalar joint (the common misnamed sprain of the ankle). Should the force continue, the tibia breaks at its junction with the medial malleolus. The radiograph is distinctive (Fig. 18). The fracture is in the sagittal plane. Starting below at the junction of the horizontal and vertical parts of the mortise it runs almost vertically upwards, emerging on the medial border of the tibia. The tip of the lateral malleolus is often avulsed but the malleolus itself does not break as a rule.

An adduction fracture is reduced by abducting the foot. (The terminology is confusing, for abduction and adduction of the foot are usually referred to as inversion and eversion.)

The *shear fracture* is not common. It is caused by a force that would elsewhere result in a crush fracture. Both malleoli break transversely at the level of the horizontal part of the mortise. The foot is displaced medially or laterally according to the direction of the force (Fig. 19). The site of the malleolar fractures makes any reduction unstable on account of the absence of side buttresses to hold the talus. In practice stability is obtained by screwing the medial malleolus to the shaft of the tibia. The medial malleolus then acts as a buffer against which the talus is pushed. The shear fracture is mimicked on occasions by a lateral-rotation fracture (Fig. 20). The essence of the shear is that both malleoli break at their junction with the shafts, and this renders closed reduction unstable.

FIG. 17
Abduction Pott's fracture.

FIG. 18
Adduction Pott's fracture.

FIG. 19
Shearing Pott's fractures. *Left:* Talus shifted laterally. *Right:* Talus shifted medially.

The *vertical fracture* is caused by longitudinal compression, and the usual history is a fall from a height. It causes either *a*) a T-shaped fracture of the lower end of the tibia with marked displacement (Fig. 21), or *b*) linear cracks running up the shaft without any displacement (Fig. 6). The latter is a trivial injury; it requires little or no treatment and recovery is speedy. The former is a serious injury and leaves a permanent disability. In view of the direction of the fracturing force skeletal traction is applied through the calcaneum. This

FIG. 20
Lateral-rotation Pott's fracture, mimicking a shearing fracture.

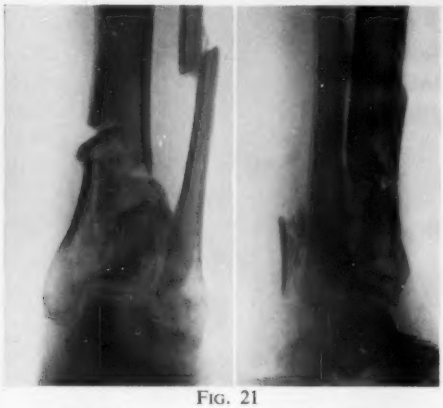

FIG. 21
Longitudinal-compression Pott's fracture.

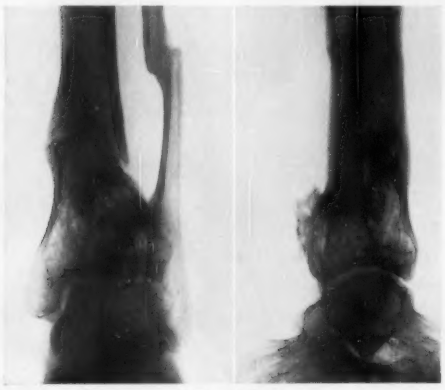

FIG. 22
The result.

is a good method of treatment because it enables the patient to start moving his ankle at once. Skeletal traction does not always produce anatomical reduction and the lateral radiograph may show a step in the articular surface of the tibia, but by moving the ankle early the patient renders congruous the opposing articular surfaces of the tibia and talus (Fig. 22). It should be noted that skeletal traction is only of use in the treatment of the vertical type of Pott's fracture. It does not, for example, reduce a lateral-rotation fracture. In order to obtain reduction of a fracture, force should be applied in a direction opposite to that causing the fracture. This is the main reason why it is helpful to work out the nature and direction of the blow. And these can be deduced from the story of the accident, the damage to the skin, and the radiographs.

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DYSPLASIA EPIPHYSIALIS HEMIMELICA

(Tarso-epiphysial Aclasis)

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This rare epiphysial dysplasia was first recognised and described as a definite entity by Trevor (1950), who collected ten cases and gave it the name of tarso-epiphysial aclasis. The only previously recorded case was that of Mouchet and Belot (1926) under the title of " tarsomégalie." Since Trevor's paper, only two further cases have been reported in the literature. One was recognised and given the title " chondrodystrophie épiphysaire " by Ingelrans and Lacheretz (1953). The medial side of the lower limb was involved in a boy of one and a half years. Both epiphyses at the knee, the medial malleolus, talus, navicular and first cuneiform were affected. The lesser trochanter, but not the femoral head, was also involved. Ossification in the centres for the navicular and first cuneiform were abnormally advanced. They regarded the condition as " a congenital dystrophy affecting the epiphyses and short bones of one ray of the limb." The second case, described by Donaldson *et al.* (1953) as " an osteochondroma of the distal femoral epiphysis," affected a girl of nine months. The inner side was involved; no other joints were radiographed, but the appearance of the knee is typical of the condition under discussion.

There is no doubt about the rarity of the condition. Enquiry at most of the orthopaedic centres in this country has produced only a total of fourteen further cases, including two in the upper limb which appear to fall into this group. The radiological appearances are sufficiently typical to make it unlikely that many examples would remain unrecognised. This communication embodies a report on this new material and a study of the total of twenty-seven cases which have now been recorded.

NOMENCLATURE

There are only three conditions so far known in which the primary defect lies in the epiphyses—dysplasia epiphysialis multiplex, dysplasia epiphysialis punctata, and the condition under discussion. It is true that the epiphyses may be involved in a number of other diseases, particularly in the Morquio-Brailsford type of chondro-osteo-dystrophy, but they are only a part, often inconstant, of the general pathological picture. It is the fate of newly recognised conditions to receive a succession of baptisms as the entity becomes better understood. There is no evidence to justify labelling this condition a dystrophy, as suggested by Ingelrans and Lacheretz, nor does the question of " rays " arise, as embryologically these are confined to the hand or foot and there is no counterpart in the more proximal segments of the limb.

Tarso-epiphysial aclasis, the name formulated by Trevor, is in the writer's view unsatisfactory for two reasons. First, although the tarsus is the most common site it is not constantly involved, even in cases affecting the lower limb. Nor need we consider the development of the tarsal bones as being basically in any way different from that of an individual epiphysis: the epiphysial plate peculiar to the latter is with very few exceptions unaffected in this condition. Furthermore it now appears that the condition is not even a monopoly of the lower limb, so that it seems to the writer no longer desirable to include the word " tarsus " in the title. Secondly, there is not simple failure of the perichondrium, where present, to model the epiphysis—that is, aclasis—but a true dysplasia or faulty growth of part of the epiphysis itself. It is true that even the unaffected side of the epiphysis is often slightly enlarged and abnormal in texture or outline. The whole upper femoral epiphysis was enlarged in Evans's case, reported by Trevor, and in Wainwright's case (Case 7 of this series)

the medial side of the talus and navicular is possibly involved although the affection was otherwise confined to the outer aspect of the limb. These are the exceptions and may be secondary manifestations. The condition, according to our present experience, however, is in general confined to the lateral or medial half of a single limb, and for these reasons the name " dysplasia epiphysialis hemimelica " is suggested.

HEREDITY AND FAMILIAL INFLUENCES

In no instance has a parent or sibling been similarly affected. It is noteworthy that the patient reported by Donaldson *et al.* (1953) was a uniovular twin, the other twin being reported as normal. " Typical exostoses " were reported present in the case of Ingelrans and Lacheretz, and a cousin of James's patient is said to have a Sprengel's shoulder, but no other associated congenital abnormalities have been noted.

Sex—Males predominate, only seven of the twenty-seven patients being girls.

Age—The youngest subject was eight months old at the times of diagnosis, the oldest twenty-three years. The majority first came under observation between the ages of two and eight.

SYMPTOMS AND SIGNS

Right and left sides are affected almost equally. The most common complaint in an otherwise healthy and normally developed child is of increasing swelling over the inner or outer aspect of knee or ankle. The swelling feels bony in consistency; the soft parts are not involved. When the inner side of the limb is affected, the mother may complain that the child is becoming knock-kneed or flat-footed in one leg, the reverse deformity being seen when the outer side of the joints is affected. The child may fall unduly frequently and there may be limitation of movement at the knee, ankle or tarsus. In one instance the knee " locked " in extension or flexion according to whether the medial ligament lay in front of or behind the prominence on the medial femoral condyle. Evans's patient, reported by Trevor, later developed painful " locking " incidents in the knee. The one adult patient also had symptoms of internal derangement of the knee.

There is occasionally pain in the ankle region, and there may be some secondary wasting of the muscles of the leg, but disability is minimal. The length of the limb is usually unaffected; in James's case (Case 4) there was a quarter of an inch of shortening, and in Brailsford's case (Case 3) there was a quarter of an inch of lengthening which later increased to one inch, part of this being due to coxa valga and a long femoral neck. Evans's patient developed half an inch of lengthening of the tibia. In one case the affected foot was markedly bigger than the other. Blood analyses have been consistently negative.

DISTRIBUTION

Characteristically the epiphysial abnormalities are confined to one side of the affected limb; the medial side was affected in nineteen, the lateral in eight, one of the latter patients (Case 7) having possibly some changes on the medial side also. In no case was more than one limb affected. Examination of the films of Mr Donald Brown's patient mentioned in Trevor's postscript (1950) as a possible bilateral case reveals what is almost certainly Köhler's disease in the right foot, but no changes suggestive of dysplasia epiphysialis hemimelica; the left knee is, however, typical. Table I shows the distribution of the lesions affecting the lower limb. It will be seen that the upper end of the femur is rarely involved, but when there are changes here there are lesions at knee, ankle and tarsus. The lower femoral epiphysis and the talus are the most common sites. With a few exceptions changes at the knee were accompanied by changes at the ankle or in the tarsus. The cuboid and the second and third cuneiforms have not so far been seen affected, and it appears probable that the involvement of the calcaneum reported in two cases has been no more than secondary distortion from the talar abnormality. The lesser trochanter was affected in two cases.

TABLE I

DISTRIBUTION OF LESIONS IN TWENTY-FIVE LOWER LIMBS

Case		Femur			Tibia		Fibula	Tarsus		
		Upper	Lesser trochanter	Lower	Upper	Lower	Lower	Talus	Navicular	First cuneiform
Mouchet and Belot								+		
Donaldson *et al.*				+						
Ingelrans and Lacheretz			+	+	+	+		+	+	+
Trevor	1						+	+		
	2			+						
	3			+			?+	+		
	4			+	+	+		+		
	5			+	+	+				
	6			+	+		+	+		
	7	+		+		+		+	+	+
	8	+	?+	+		+		+	+	+
	9					+		+	+	
	10			+						+
T. J. F.	1			+	+					+
	2						+	+		
	3	+		+	+		+	+		
	4					+		+		
	5						+	+		
	6			+						
	7			+			+	+	?+	
	8							+		
	9			+	+	+		+	+	+
	10					+		+	+	+
	11			+		+				+
	12					+		+		

RADIOLOGICAL APPEARANCES

These are characteristic and have been admirably described by Trevor. Except in the case of the smaller tarsal bones, one side only of the epiphysis is involved. There may be generalised though irregular enlargement of the affected side, or more commonly there can be seen a number of separate centres of ossification, irregular in shape and size, discrete from the main epiphysis, and often so closely packed together that they appear to form a single mass of bone. From the lower femoral epiphysis they usually project sideways, downwards and backwards, but occasionally they lie in front of the coronal plane. As Trevor pointed out, one may see at the knee an anterior projection from the femoral epiphysis and posterior

projection from the tibia, or vice versa. If the projection impinges on the opposing articular surface, varus or valgus deformity is produced, but large masses may be seen on radiographs projecting from the side and margin of the epiphysis without causing any angular deformity whatever. In young subjects these secondary centres are often widely separated from their parent epiphyses, but with the passage of time they tend to fuse with each other and then with the main body of the epiphysis, although this does not always occur (Case 9). Secondary deformation in the opposing epiphysis at a joint is not uncommon.

The metaphysis is in general unaffected, but in two cases there was considerable widening of the femoral neck with, in one instance, elongation of the neck and coxa valga; and in two examples changes in the lower fibular epiphysis were associated with quite marked widening of the metaphysial region. The length of the bones is usually unaltered, but exceptions have already been quoted. No bowing of the shaft of a long bone has been seen, except in the ulna in Case 13, which also underwent progressive shortening.

When the tarsus is involved there may be massive enlargement of the body of the talus, the articular surface of which sometimes appears crenated in the antero-posterior view, and the whole bone may be tilted to one side or the other. Even when the whole body is enlarged it is obvious that one side is much more severely affected than the other. There is little or no diastasis of the tibio-fibular mortise. The head of the talus is less often involved than the body, but it was noteworthy that in several cases the head had lost its normal rounded contour and had become almost square in outline on the lateral view. The navicular and first cuneiform are commonly included in the process, but not the other tarsal bones. In cases seen early enough there is premature appearance of the bony centres for the tarsal bones, particularly when the medial side of the foot is involved, and these centres remain larger than their fellows in the opposite foot for several years. Slight increase in length of the medial metatarsals has also been observed. These last two points were considered of some importance in placing Freebody's case (Case 12) into the group, as there was enlargement of the centres for both multangular bones and lengthening of the second metacarpal. The upper fibular epiphysis and that for the patella have not been seen involved.

The whole skeleton has been radiographed in many of the cases reported here, and no abnormalities elsewhere have been detected.

PROGRESS

The swelling increases gradually for the first few years of life, and varus or valgus deformity at a joint may appear or increase during this period. After the age of six or seven there may be little deterioration, the swelling increasing only *pari passu* with the general growth of the child. Radiologically new centres of ossification may appear, and the multiple bony centres increase in size and tend to fuse together into a common mass which itself usually fuses with the rest of the epiphysis, where changes in texture in the affected region overshadow the previously clear-cut division between the normal and abnormal epiphysial bone. When the projection is a long one and movements put a strain on its junction with the rest of the epiphysis it appears that sometimes part or all of the projection is broken off, union of the fractured cartilage fails to occur, and a fragment remains permanently separated from the rest of the epiphysis by attenuated fibrous tissue or even a false joint. No elderly patients have yet come under observation but one must presume that many of the misshapen joints will be liable to later degenerative arthritis.

TREATMENT

Part or all of the outgrowth has been removed in a number of patients with benefit in every case. When there had been valgus or varus deformity from the pressure of the outgrowth this was improved or completely corrected. Apart from cosmetic interventions, this appears to be the main indication for operation. Although post-operative radiographs often show that a considerable proportion of the affected part of the epiphysis has been left untouched

no ill-effects have resulted, no attempt at regeneration of the excised portion takes place, and normal and remaining abnormal parts of the epiphysis continue to grow as if there had been no intervention. There are, however, many cases, particularly those involving the body of the talus, in which it is obvious that any corrective procedure would involve a prohibitively large removal of the bone. On two occasions adequate removal of the outgrowth at the knee so damaged the attachments of the semilunar cartilage that it had to be excised. At the knee a supracondylar osteotomy was done to correct genu valgum in two patients.

PATHOLOGY

At operation the outgrowth is found to be covered with glistening bluish cartilage similar to that of the normal epiphysis. Its surface may be smooth and its line of junction to the rest of the epiphysis indistinguishable. More often, however, its surface is irregular and slightly knobbly and a groove of varying depth demarcates it from the normal tissue. When a fragment has become entirely separated from the rest of the epiphysis it is found lying snugly fitting a bed lined with cartilage, to which it is attached by bands of fibrous tissue. In some areas movement has produced the appearance of a false joint.

Microscopically the appearances are like those of an osteochondroma. There is a thick zone of hyperplastic cartilage distinguishable from that of the rest of the epiphysis only by some irregularity of cell size and distribution. Active normal enchondral ossification is taking place around the bony centres, which are irregular in size and shape and may be rather deficient in marrow spaces, making the bone relatively dense. The bone itself shows otherwise no abnormality. There is therefore apparently no characteristic feature on which a diagnosis can be made from biopsy material.

DIAGNOSIS

While one may exceptionally detect so rare a condition clinically, diagnosis from radiographs is usually simple. The restriction of changes to those epiphyses or parts of epiphyses lying on one side of a single limb, the characteristics of those changes, and the absence of abnormalities in other parts of the skeleton permit no alternative diagnosis. The other epiphysial dysplasias are different in a host of ways. Synovial osteochondromatosis may produce somewhat similar radiological appearances, but the patient is older and the epiphysial regions are unaffected. Cases have been seen in which a single bony outgrowth from some tarsal bone caused suspicion of the condition, but in the present state of our knowledge it has been considered wiser not to include them in this group, although they may be closely related.

ETIOLOGY

It is fascinating, though perhaps profitless, to delve into the possible causes of this condition. We can be fairly sure that the cause, whatever it may be, is active early in the fifth week of foetal development. The limb bud for the arm first appears on the twenty-sixth day, for the leg on the twenty-eighth day. We know (Duraiswami 1952) that insulin and a number of other substances can produce multiple abnormalities of the limbs which vary in distribution largely according to the exact time at which they are introduced into a chick embryo. Most of the experimental work on the production of monomelic abnormalities has been done on chicks or on amphibia, but there is reason to believe that we can safely apply the results of such experiments to mammalia. Saunders (1948) has shown how the orderly sequence in which the parts of the chick's wing develop proximo-distally is controlled by the apical ectoderm of the limb bud; extirpation of this apical cap suppresses further distal development of the wing. If it is done on the third day the entire wing is suppressed. If extirpation is delayed for a few hours, growth of the humerus proceeds normally but nothing distal to it, and so on. Within a single day, even though the limb has grown little in size, it has developed all the basic cells destined to form the entire limb, and damage to the apical ectoderm after this stage will not prevent the normal development of the limb. When the upper limb apical

growth zone with its ectodermal cap is grafted to the stump of a leg bud typical wing parts are formed appended to the proximal leg structures. If only the pre-axial part of the apex of the early wing bud is extirpated a wing develops lacking the pre-axial element of the humerus, and minus a radius and the radial half of the carpus, metacarpus and phalanges. Agenesis of the ulnar side of the wing follows removal of the post-axial half of the apical cap. Tschumi (1954), working with Xenopus larvae (a variety of toad), investigated the effect of local application of dilute nitrogen mustards to the limb bud and produced various growth abnormalities, particularly in the metacarpus.

Streeter (1930), in his work on intra-uterine amputations and other developmental abnormalities of the limbs, concluded that all are due to some local defect in the limb bud itself.

In dysplasia epiphysialis hemimelica we have no knowledge of what the primary fault may be, whether in the original germ-plasm, the " organisers " within the foetus, focal haemorrhages, " bleb " formation or any other of the many etiological factors that have been suggested; it may even be due to some cause outside the embryo itself. But we can be fairly sure that it affects either the pre-axial or post-axial part of the apical cap of a single limb bud, probably for a very short time and in a minute area, and that the localisation of the future lesions depends on the exact timing and site of this fault.

I am deeply indebted to all those who have so generously loaned the clinical notes and radiographs of their cases reported here, and to the many others who kindly searched their records for further examples although without avail. My special thanks are also due to Mr David Trevor and to those whose cases he reported in 1950 for making available to me the follow-up material on which the notes on " Progress " and " Treatment " were largely based. I am most grateful, too, to Professor J. D. Boyd and Dr P. Tschumi of the University Department of Anatomy at Cambridge for their willing assistance and advice on recent embryological developments, to Dr A. M. Barrett of the Department of Pathology for the photomicrographs and pathological reports, and to Miss Westoby at Addenbrooke's Hospital for the photographic work; and to Mr J. N. Aston for the operation sketch of Case 9. Finally I offer profound tribute and thanks to my father for his inspiration, his zest, and his unstinted guidance and help.

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APPENDIX

DETAILS OF THIRTEEN COLLECTED CASES OF DYSPLASIA EPIPHYSIALIS HEMIMELICA

Case 1 (reported by courtesy of Mr A. L. Eyre-Brook)—Girl aged two years three months. Six months' history of frequent falls, with deformity of the right knee. There was bilateral genu valgum, more marked on the right. At operation the infero-posterior part of the medial femoral condyle was lumpy and rather bluer than the rest of the epiphysis. The greater part was cut away, correcting the genu valgum. The medial semilunar cartilage required removal. A skeletal survey showed lesions only in the medial side of both epiphyses at the knee and in the right first cuneiform (Figs. 1 and 2). Radiographs of the specimen are shown in Figure 3.

Case 2 (reported by courtesy of Mr D. Freebody)—Boy aged two years nine months. There had been pain and swelling of the outer side of the left ankle for one month after a fall. Similar trouble for a

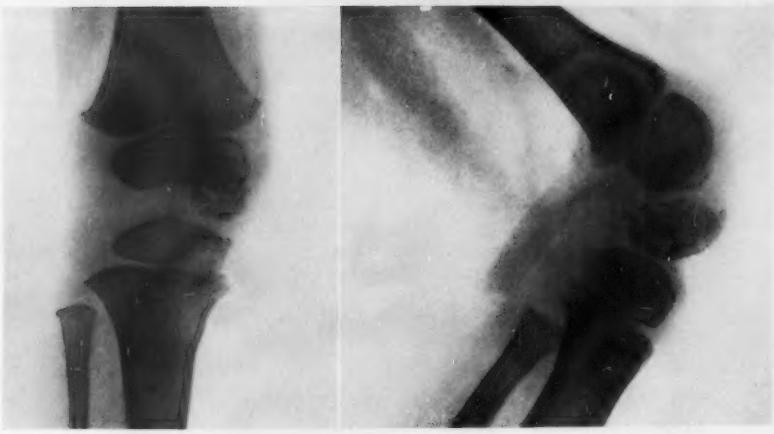

FIG. 1
Case 1—Right knee. The medial sides of both epiphyses are involved.

FIG. 2
Case 1—Feet. The right shows enlarged first cuneiform with multiple centres of ossification.

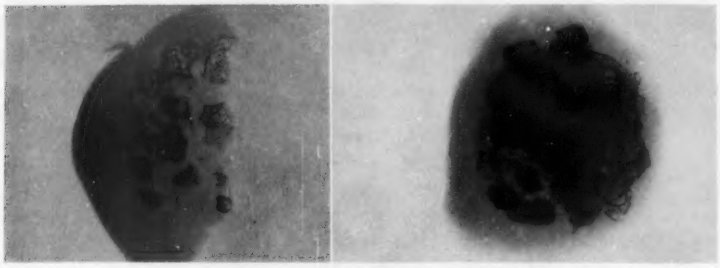

FIG. 3
Case 1—The specimen from the medial femoral condyle.

few weeks one year previously, not associated with injury. Eversion of the foot, which was otherwise fully mobile, was blocked by a bony prominence below and in front of the lateral malleolus. Radiographs showed involvement of the outer side of the talus and of the inferior fibular epiphysis, the metaphysis of which was splayed out (Fig. 4). The centre for the navicular was much larger than in the right foot. At operation two bony fragments were found attached by fibrous tissue to the lateral aspect of the talus, one projecting into the sinus tarsi. They were removed, together with a third mass of cartilage and bone arising from the medial aspect of the lower fibular epiphysis. The child made an excellent recovery.

Case 3 (reported by courtesy of Dr J. F. Brailsford)—Boy aged five. Bowing of the right leg with a quarter-inch of lengthening. In the course of two years three months the shortening increased to one inch. Radiographs (Figs. 5 to 7) showed irregular enlargement of the lateral half of the femoral head, coxa valga and a long femoral neck, a multi-centred mass growing from the outer side of the lower femoral epiphysis, and slight changes in the upper tibial epiphysis laterally. At the ankle there was an outgrowth from the outer side of the body of the talus and a large mass of bone arising from the fibular epiphysis, causing considerable diastasis.

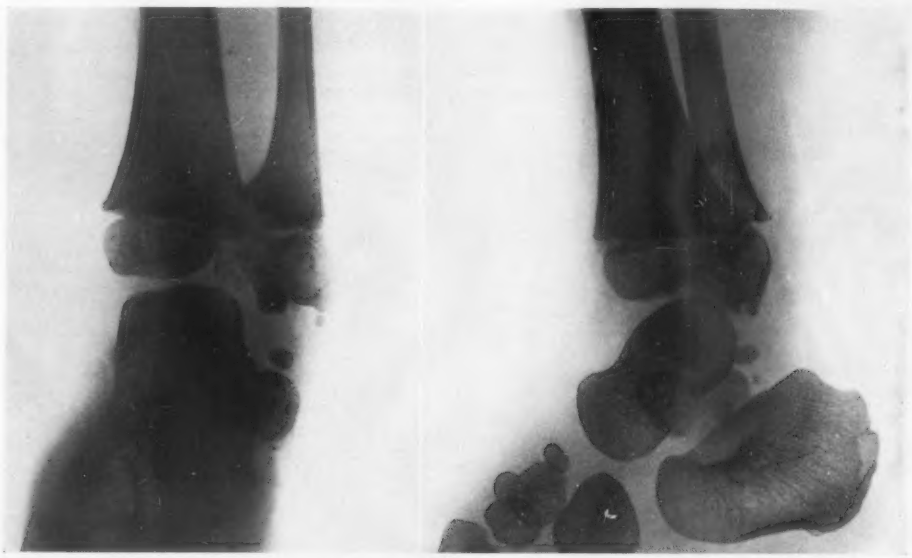

FIG. 4

Case 2—Left ankle, showing lesions of talus and fibular epiphysis, with splaying of the fibular metaphysis.

Case 4 (reported by courtesy of Mr J. I. P. James)—Boy aged eight years six months. At the age of eighteen months is said to have fractured his right ankle. Had been under observation elsewhere for the last three years and an operation performed to remove a bony prominence. The right medial malleolus was still extremely prominent, the ankle displaced laterally and the foot in slight valgus. Dorsiflexion was limited to 90 degrees. The right leg was underdeveloped and a quarter-inch shorter than the left. Hip and knee were normal. Radiographs (Fig. 8) showed irregular enlargement of the medial side of the talus, which projected forwards and also upwards into the medial half of the tibial epiphysis, where there appeared to be a secondary concavity. The medial malleolus was enlarged, and the navicular rather deeper than in the left foot. The patient has been kept under observation for five years, during which there has been little deterioration, the swelling increasing only *pari passu* with the growth of the child and the degree of valgus only increasing slightly (Fig. 9).

Case 5 (reported by courtesy of Mr K. I. Nissen)—Girl aged six years six months. History of painless swelling of the outer side of the left ankle for six months. There was a bony prominence in front of the outer part of the ankle, particularly in plantar flexion. There was some limitation of ankle and subtalar movements. Radiographs (Fig. 10) showed irregular enlargement of the outer half of the body of the talus, the head of which was flattened. The lower fibular epiphysis was elongated and bulbous in its distal part. Other joints were normal.

FIG. 5

Case 3—The outer part of the right upper femoral epiphysis is irregularly enlarged, and the femoral neck long, thick and valgus.

FIG. 6

FIG. 7

Case 3. Figure 6—Right knee. The outer part of the tibial epiphysis is minimally affected. Figure 7—Right ankle. Talus and fibular epiphysis are involved, upward growth from the latter apparently producing diastasis.

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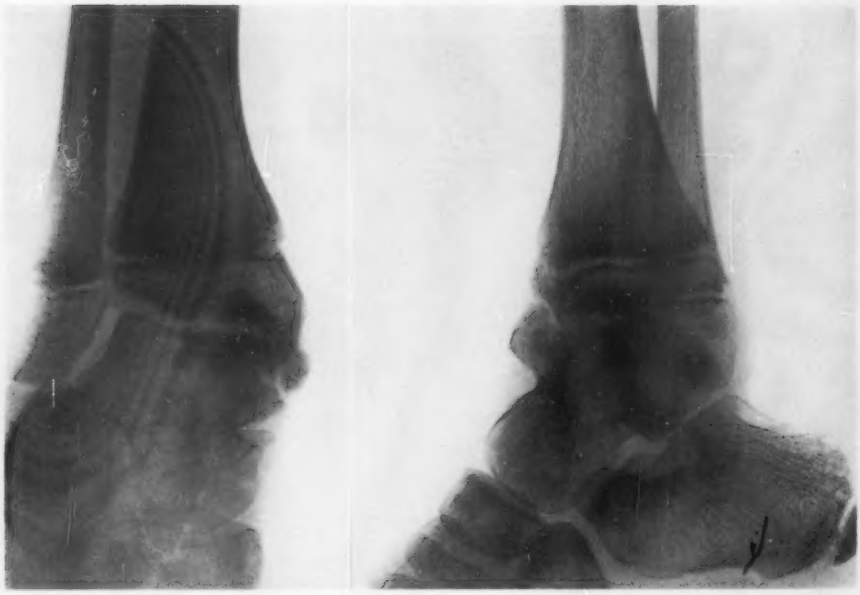

FIG. 8
Case 4—Right ankle. The outgrowth from the talus has produced a cavity in the tibial epiphysis, which is itself affected.

FIG. 9
Case 4—The same ankle four years later.

Case 6 (reported by courtesy of Mr David Trevor)—Boy aged three. Painless swelling of the inner side of the right knee noticed since a minor fall ten days previously. The swelling was bony hard; there was no restriction of movement. At birth he was said to have had a lateral rotation deformity of the right leg; this recovered spontaneously in six months. A cousin is said to have a Sprengel's shoulder. The swelling increased and genu valgum developed, and one year three months later the

FIG. 10

Case 5—The ankle, fibula and outer part of talus involved. Note the flattening of the head of the talus.

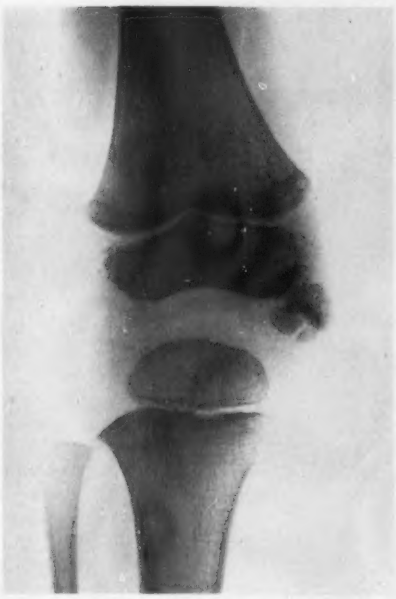

FIG. 11

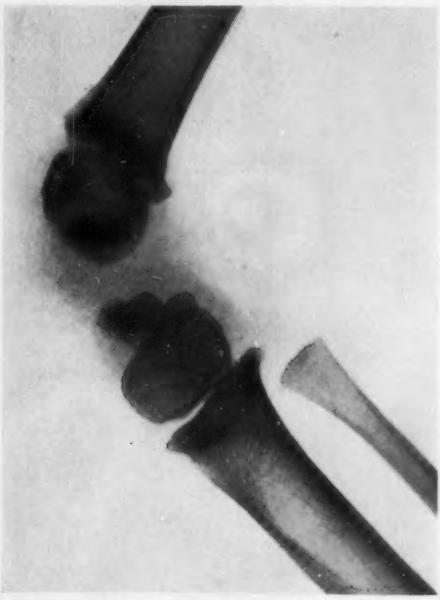

FIG. 12

Case 6. Figure 11—Involvement of the medial side of the femoral epiphysis. Figure 12—An earlier lateral view showing the wide separation of the new centres from the parent epiphysis.

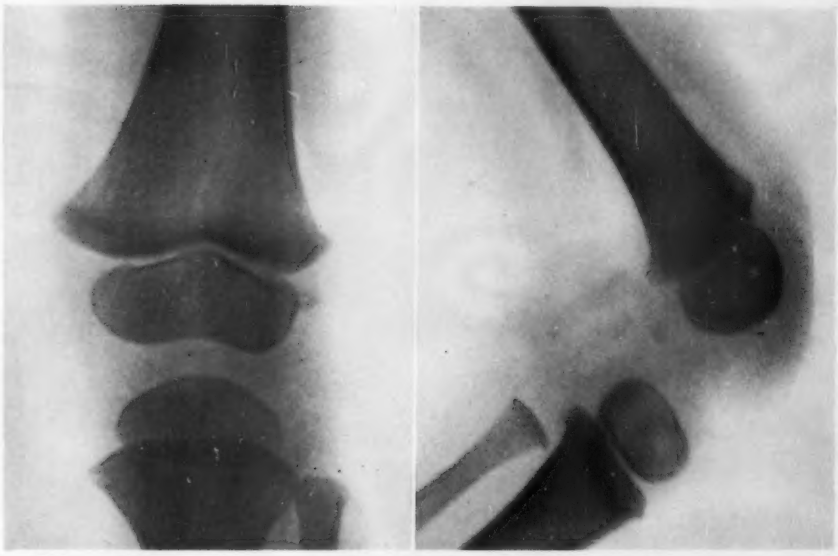

FIG. 13
Case 7—The knee, aged one year. A new centre is just visible behind the lateral femoral condyle.

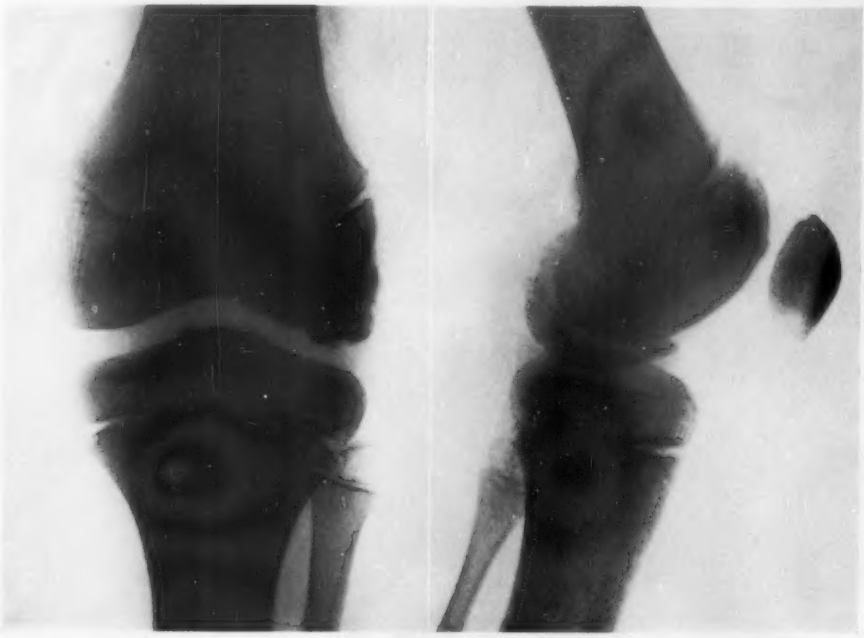

FIG. 14
Case 7—The same, nine years later.

greater part of the outgrowth was removed. Radiographs (Figs. 11 and 12) showed at least three separate centres of ossification lying below and behind the medial side of the femoral epiphysis. No other joint was affected.

Case 7 (reported by courtesy of Mr Denys Wainwright)—Boy aged ten. When one year old " went off his feet " for three weeks. Radiographs (Fig. 13) showed a small centre of ossification just visible behind and lateral to the outer side of the left lower femoral epiphysis. Attended aged ten complaining of pain in the left ankle which was thickened around the lateral malleolus; there was marked rigidity of the foot. Radiographs (Fig. 15) showed a bony mass below and in front of the lateral malleolus and another projecting from the back of the body of the talus. The head of the talus was flattened and the tubercle of the navicular was enlarged and irregular; this however may be analogous to a large os tibiale externum. The outer posterior part of the lower femoral epiphysis was now obviously involved (Fig. 14). The outgrowth from the lateral malleolus was removed.

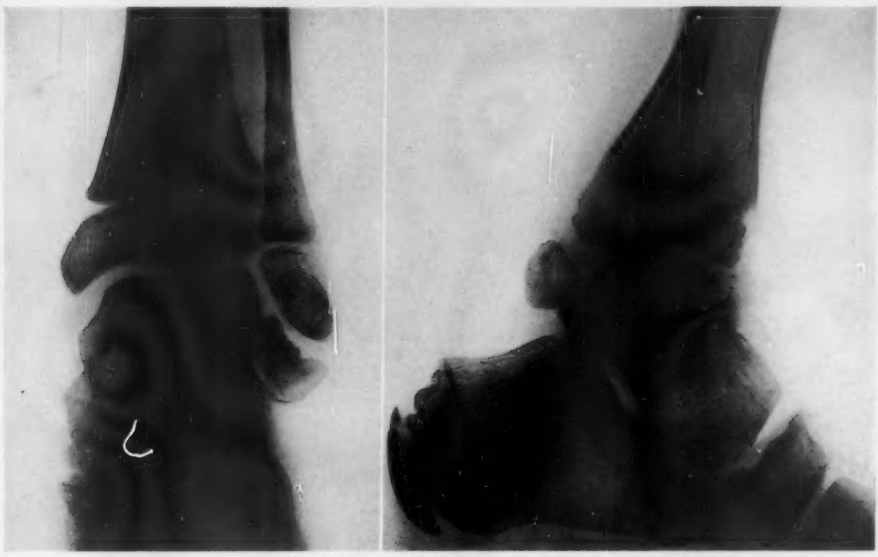

FIG. 15

Case 7—The ankle. Talus and lateral malleolus are involved; the head of the talus is flattened; the tubercle of the navicular is enlarged and irregular.

Case 8 (reported by courtesy of Mr F. C. Dwyer)—Boy aged five. Presented with valgus feet, limited inversion of the right foot and wasting of the right calf muscles. Radiographs (Fig. 16) showed an irregular outgrowth from the head and neck of the talus, a large ossification centre for the navicular (that on the left foot had not yet appeared) and slight lengthening of first and second metatarsals. Removal of the " exostosis " led to almost full recovery of the range of inversion.

Case 9 (reported by courtesy of Mr D. Ll. Griffiths)—Man aged twenty-three. He complained that for nine years his right knee had been increasingly liable to give way and lock in about 60 degrees of flexion, when he could feel a lump antero-medially. With rest this went down and movement recovered. Ten years previously he had had an operation on the right foot which had always " turned out "; this had relieved him. On examination the whole right lower limb was wasted. The medial tibial condyle was prominent. The knee joint was crepitant and lacked full extension; no rotary movement was possible. The foot was held in marked valgus. Ankle and tarsal movements were severely restricted; the talus was prominent in front of the medial malleolus. Radiographs (Fig. 17) showed that both epiphyses on the medial aspect of the knee had been involved and a large irregular mass of bone lay antero-medially between the joint margins. In the foot and ankle (Figs. 18 to 20) there were severe changes in the medial malleolus, medial side of the talus, the navicular and possibly the first cuneiform. The knee was explored by dividing the medial ligament. Large bony outgrowths were removed from femur and tibia, the greater part of those from the latter, which was in two portions,

FIG. 16
Case 8—Right foot shows lesion of head of talus, large navicular ossification centre and slight lengthening of first and second metatarsals. Left foot shown for comparison.

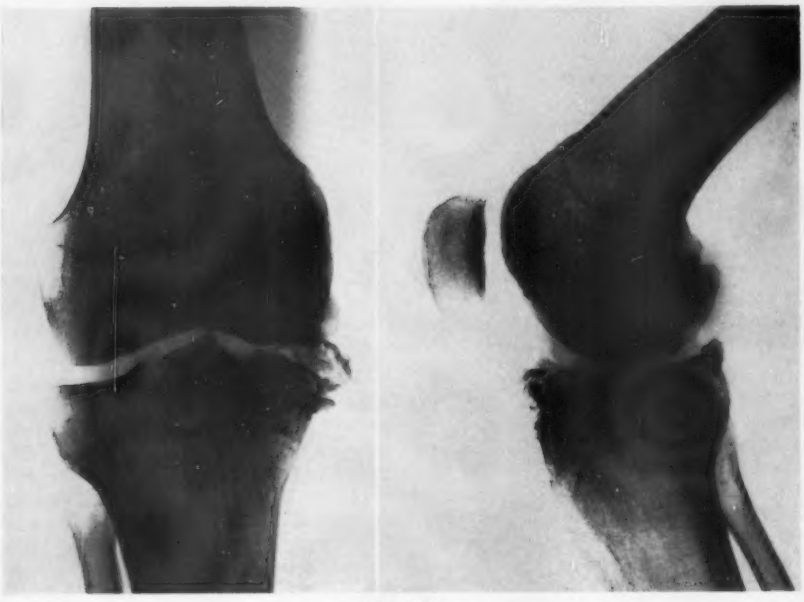

FIG. 17
Case 9—Right knee. The inner sides of both femur and tibia are involved.

FIG. 18

FIG. 19

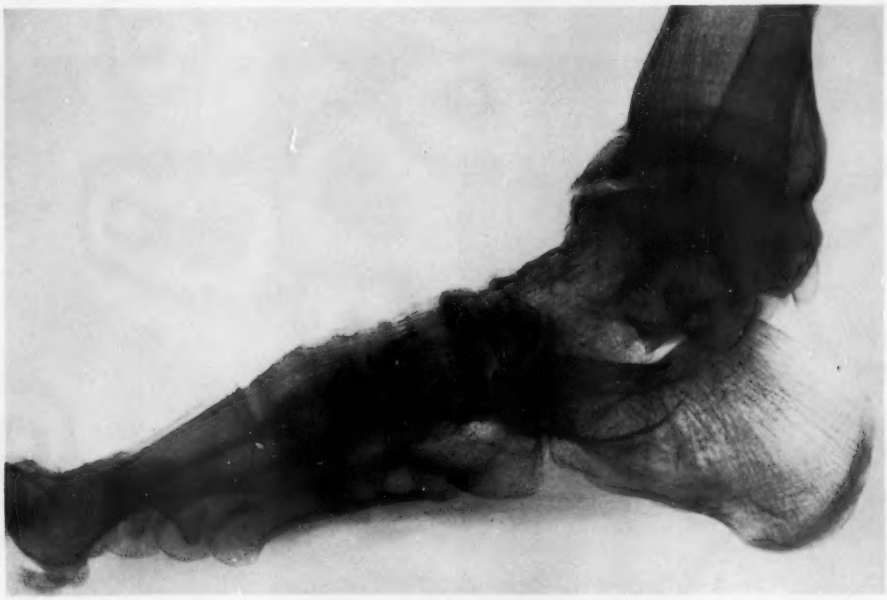

FIG. 20

Case 9—Right ankle and foot. The medial malleolus, talus, navicular and possibly the first cuneiform are affected.

FIG. 21

Case 9—Operative findings. The outgrowth from the femur has been removed. The medial cartilage and tibial mass are still in situ.

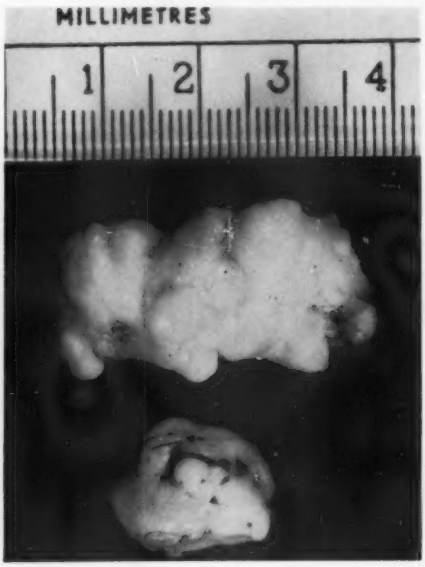

FIG. 22

FIG. 23

Case 9. Figure 22—The specimen from the tibia. Figure 23—Radiograph of the specimen.

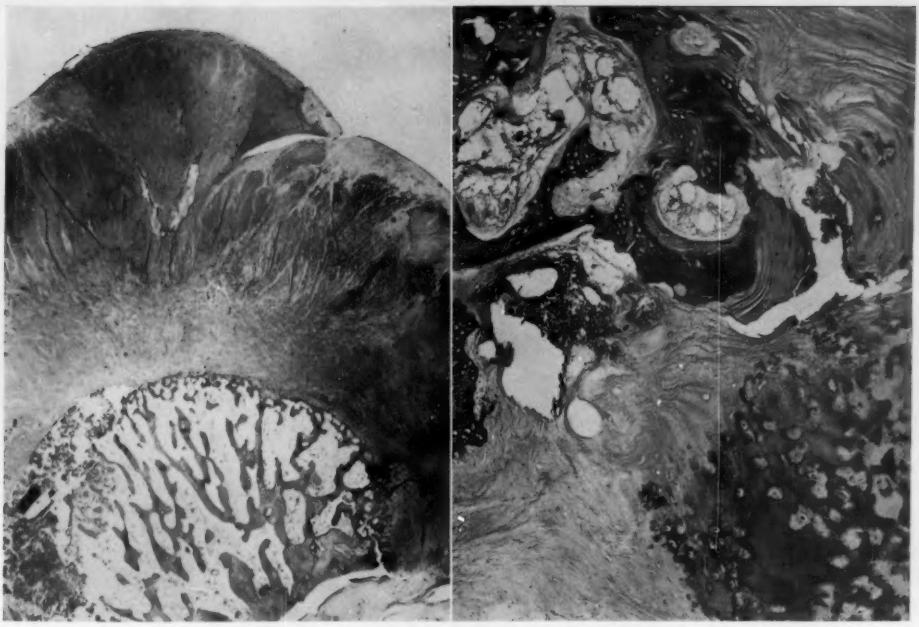

FIG. 24 FIG. 25

Case 9—Photomicrographs of tibial specimens. For description see text. (Fig. 24, ×10; Fig. 25, ×75.)

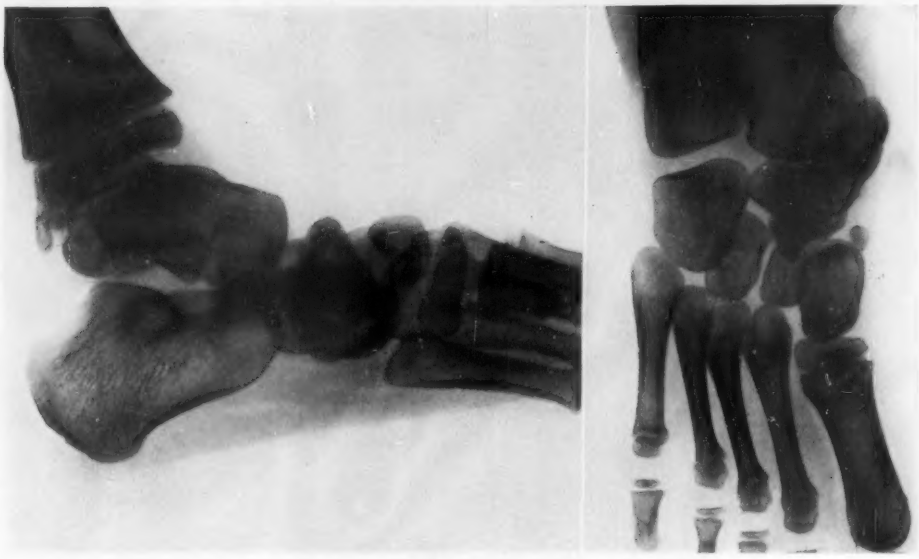

FIG. 26

Case 10—Right foot with lesions of medial malleolus, talus, navicular, and first cuneiform.

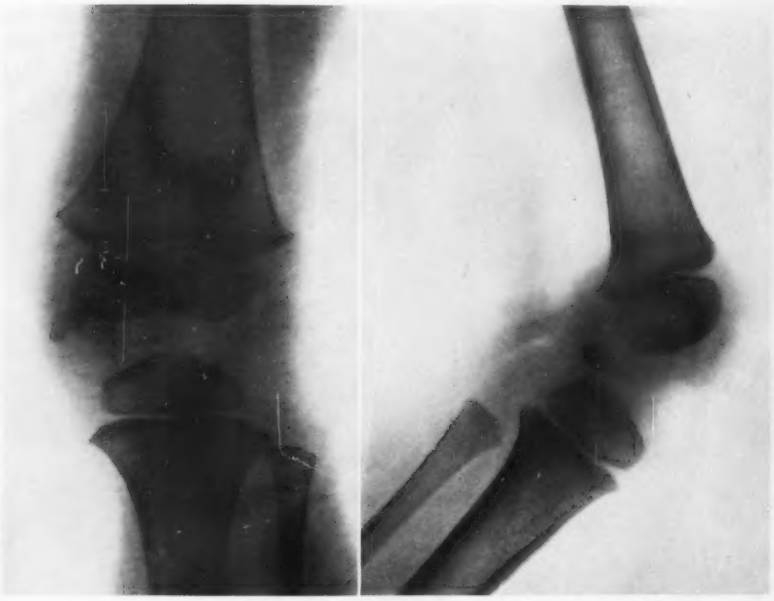

FIG. 27

Case 11—The knee with typical outgrowths from the medial side of the femoral epiphysis.

FIG. 28

FIG. 29

Case 11. Figure 28—The ankle showing involvement of the medial malleolus. Figure 29—The foot with multiple centres for the first cuneiform.

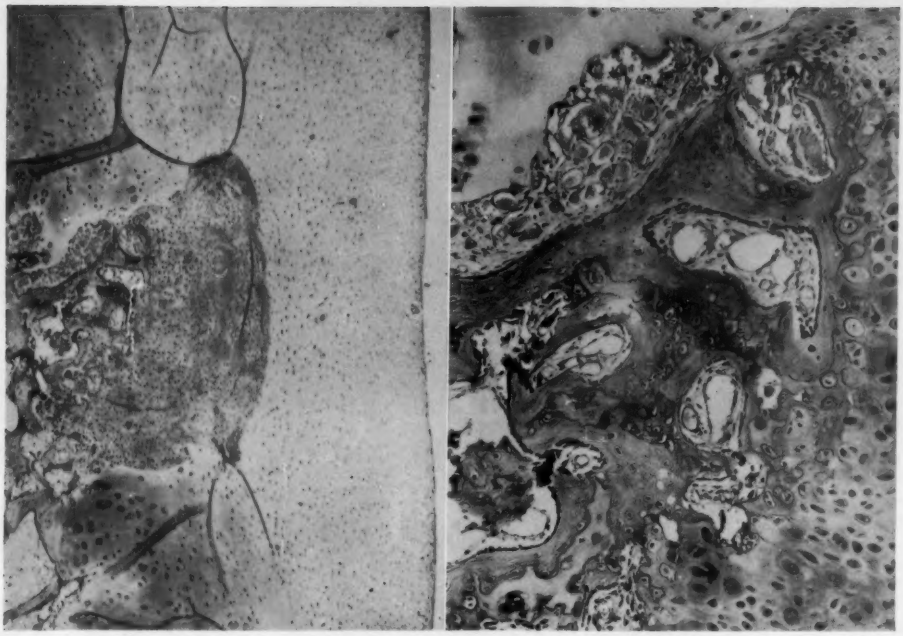

FIG. 30 FIG. 31

Case 11. Figure 30—Photomicrograph of specimen from femur. (×25.) Figure 31—The same. (×90.)

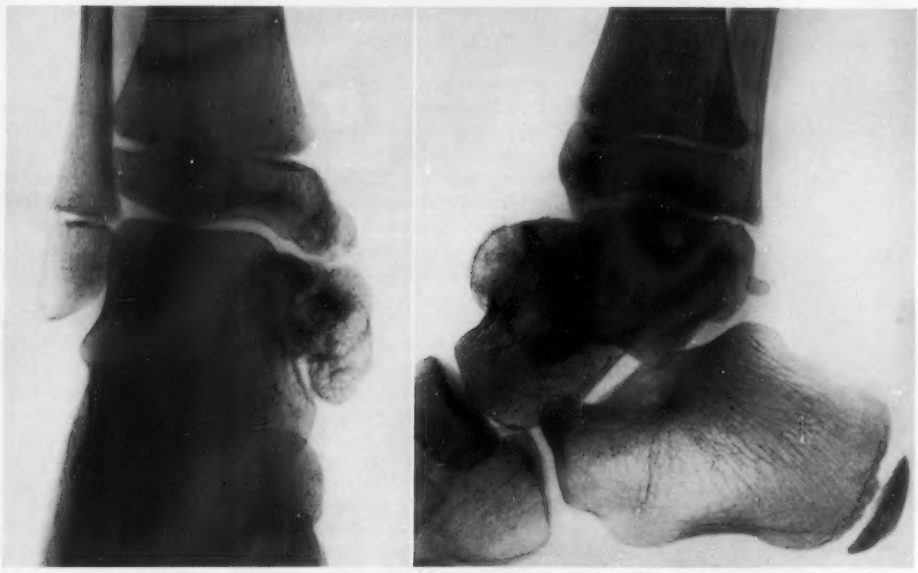

FIG. 32

Case 12—Right ankle showing outgrowths from talus and medial malleolus.

FIG. 33
Case 13—Right wrist showing enlarged distorted scaphoid. Enlarged centres for lunate, trapezium, trapezoid and first metacarpal epiphysis. Left wrist shown for comparison.

lying almost detached in a bed they had made for themselves (Figs. 21 to 23). The semilunar cartilage was excised. Photomicrographs are shown in Figures 24 and 25. Histology showed that all of the bone and much of the cartilage had undergone necrosis, presumably due to deprivation of blood supply when detachment occurred. Degenerative changes of various kinds were present in the surviving cartilage: near one surface the fibrillated appearance characteristic of osteoarthritis was seen. The bone was in the form of irregularly round islands which could represent centres of ossification. There was no evidence that ossification was still progressing when necrosis occurred.

FIG. 34
Case 13—The same eight years later. Elongation of second metacarpal.

Case 10 (reported by courtesy of Mr H. L.-C. Wood)—Girl aged eight. Lump on right ankle noticed first at age of three, slowly increasing since. Limitation of ankle and tarsal movements. A lump could be felt behind the medial malleolus and also on the outer side. She stood with the foot in valgus. There was an inch of wasting of thigh and calf; no inequality in length of limbs. Radiographs showed involvement of medial malleolus, talus, navicular and first cuneiform (Fig. 26).

Case 11 (under care of the author)—Boy aged two years nine months. Six months' history of walking with a stiff left leg, with difficulty in both bending and straightening the knee. With a little force both movements could be performed, in each case with an audible snap on the inner side. The medial femoral condyle was unduly prominent and there was slight genu valgum, equal on the two sides. On exploration the snap was found due to the posterior margin of the medial ligament riding across the protuberance, the removal of which resulted in normal knee movements. Figures 27 to 29 show the lesions in the medial femoral condyle, the medial malleolus and the first cuneiform. There was very slight enlargement of the epiphysis of the first metatarsal. Sections of the specimen (Figs. 30 and 31) showed trabecular bone, the marrow spaces of which were filled with fibro-fatty

FIG. 35
Case 14—Right wrist showing a deep misshapen ulnar epiphysis, apparently fused laterally, a short bowed ulna and a large irregular lunate affected mainly on its ulnar side.

tissue, covered by a cap of hyaline cartilage, the cells of which were variable in size and were spaced irregularly in clumps.

Case 12 (reported by courtesy of Mr F. C. Dwyer)—Girl aged five. Complained of a lump on the upper and inner surface of the right foot. This increased gradually in size. At the age of eight a bony mass was removed from the talus and the medial malleolus was trimmed. She now walks perfectly normally and there has been no recurrence of the swelling four years later. Radiographs (Fig. 32) showed involvement of the back and front of the medial side of the body of the talus, and enlargement of the medial malleolus.

Case 13 (reported by courtesy of Mr D. Freebody)—Boy aged seven. History of restriction of movement of the right wrist with painless swelling on the radial side increasing gradually over several months. No radial deviation was possible; flexion and extension were slightly limited. Radiographs (Fig. 33) showed a grossly enlarged and distorted scaphoid, the centre for the trapezium enlarged and misshapen, and slight enlargement of those for the lunate, trapezoid and first metacarpal. The patient was kept under observation. Swelling, restriction of movement and weakness gradually increased. Figure 34 shows the appearance eight years later; the second metacarpal was unduly long and massive. At this time the proximal two-thirds of the scaphoid were resected, together with a second fragment apparently arising from the lunate, to which it was attached by fibrous tissue. The wrist made an excellent recovery, with full power, no disability, and only slight restriction of movement.

Case 14 (reported by courtesy of Mr F. C. Dwyer)—Girl aged twelve. First seen aged three because of undue prominence of the right lower ulnar epiphysis. With time the swelling became gradually less conspicuous and she suffers no disability. Radiographs (Fig. 35) showed a short bowed ulna with a deep misshapen lower epiphysis which had apparently fused to the metaphysis on the radial side, a fact presumably responsible for the under-growth and distortion of the shaft. The lunate was enlarged and irregular in texture, the ulnar side being chiefly affected.

PRIMARY RETICULO-SARCOMA OF BONE

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Although Oberling and Raileanu (1932), after an extensive study of the histology of Ewing's tumour, concluded that it was to be regarded as a reticulo-sarcoma arising in the bone marrow it was not until 1939 that Parker and Jackson published the first paper on this subject recording seventeen cases collected from their own material, and from the Registry of Bone Sarcomas. From that time there have been numerous papers in the American literature recording further series of cases, but British publications have been limited to a paper by Valls *et al.* (1952) and one case reported by Fripp and Sissons (1954). This paper is concerned with reporting seven cases of reticulo-sarcoma of bone and considering the relationship between this tumour and Ewing's sarcoma.

In 1921 Ewing described a tumour which he called a diffuse endothelioma of bone. It occurred particularly in children, was often multifocal in the skeleton and rapidly fatal with generalised metastases. This paper was followed, from then until now, by a spate of papers on the subject, many of them describing isolated cases with inadequate pathological reports and usually no necropsy records. In 1933 Colville and Willis published the first critical paper on the subject. They reported the case of a child, found to have a neuroblastoma at necropsy, which presented in life the clinical picture of Ewing's syndrome. Willis (1940) reported a similar case and suggested that it was very likely that all cases of Ewing's tumour could be explained in this way. Although there can be no doubt that this view was true of many of the published cases the passage of time and the publication of series of cases in the American literature with adequate necropsy reports makes it clear that there is a primary tumour of bone which many still call Ewing's tumour. Willis (1953), while rightly pointing out that many of the cases presenting the clinical and pathological picture of Ewing's syndrome are metastatic neuroblastomas if occurring in children and adolescents, and metastatic carcinomas if occurring in adults, accepted the fact that primary reticulo-sarcoma of bone accounts for a minority of such cases.

During the past ten years there have been several important reviews of Ewing's tumour in the American literature. Lichtenstein and Jaffe (1947) reported seventeen cases, four of which came to necropsy. Coley *et al.* (1948) based their study on ninety-one cases, and Geschickter and Copeland (1949) reviewed a series of 167 cases with necropsy reports on twelve. McCormack *et al.* (1952) reviewed all the bone tumours seen at the Mayo clinic between 1907 and 1949. They found eighty cases which they regarded as Ewing's tumour. In all these papers Ewing's tumour is regarded as an entity distinct from reticulo-sarcoma of bone.

Since Parker and Jackson's paper in 1939 there have been a number of papers reviewing large series of cases of reticulo-sarcoma of bone. Ivins and Dahlin (1953) reviewed all the available material in the period 1905–52. They found forty-nine cases of reticulo-sarcoma of bone and over eighty cases of Ewing's tumour. Of the forty-nine cases of reticulo-sarcoma thirty-two had already been reported by McCormack *et al.* (1952). Francis *et al.* (1954) reported forty-four cases of reticulo-sarcoma of bone and smaller series have been recorded by Simmons (1939), Chan Szutu and Chih-Kuang Hsieh (1942), Sherman and Snyder (1947), Khanolkar (1948), Uehlinger *et al.* (1948) and Fruhling *et al.* (1954).

Although in most papers in the American literature Ewing's tumour and reticulo-sarcoma are regarded as separate entities, there has been a tendency in recent years to stress the difficulty of separating them, and Geschickter and Copeland (1949) described reticulo-sarcoma of bone

as atypical Ewing's sarcoma. In the case of Ewing's tumour the main features are said to be the youth of the patient, the rather characteristic radiographic picture, the radiosensitivity of the tumour, the very bad prognosis, and the uniform histological appearance. In the case of reticulo-sarcoma of bone the main features are their occurrence in an older age group, the relatively good prognosis, and the pleomorphic histological picture with the emphasis on the presence of reticulin fibre between the tumour cells (Table I).

TABLE I

SUMMARY OF MAIN DIFFERENCES BETWEEN EWING'S TUMOUR AND RETICULO-SARCOMA OF BONE

	Ewing's sarcoma	Reticulo-sarcoma of bone
Age	Early life (4½–25 years)	Later life (20–40 years). Extremes 10–59 (Parker and Jackson)
Sex	Male : Female = 2 : 1	Male : Female = 2 : 1
Site	Shaft of long bones. 40 per cent in femur or tibia	Flat bones, trunk, metaphysis of long bones extending to shaft
Onset	6 weeks to 2 years; average 9 months (McCormack *et al.*)	6 months to 8 years; average 1–2 years
First symptom	Pain or swelling	Pain or swelling
Fever	Present in 30 per cent	Infrequent
Radiology	Not diagnostic: early stages new bone formed—later stages osteolytic	Not diagnostic: usually osteolytic
Radiotherapy	Sensitive—reappearance of normal-looking bone	Sensitive—forms more dense bone after treatment
Prognosis	Poor. Five-year survival 10–15 per cent	More favourable. Five-year survival 35–45 per cent
Gross specimen	Bulk of tumour subperiosteal; often reactive new bone in medulla	Tends to be localised in area of metaphysis
Microscopy	Uniform cell masses separated by trabeculae of collagen. No reticulin fibre in cell masses	More pleomorphic histology. Reticulin fibre between cells

Oberling (1928) and Oberling and Raileanu (1932) went exhaustively into the histogenesis of Ewing's tumour and concluded that it was a reticulo-sarcoma with a variable histological picture similar to that seen in reticulo-sarcomas arising in lymph nodes. Sternberg (1935) regarded it as an undifferentiated sarcoma, and Stout (1943), in an excellent critical paper on the subject, found himself unable to separate the two tumours on histological grounds. Willis (1953) accepted the existence of a reticulo-sarcoma of bone and stated: " whether there does indeed exist a primary tumour of bone, other than reticulum-cell sarcoma, capable of giving Ewing's syndrome is, I think, still uncertain." Lichtenstein and Jaffe (1947) and Lichtenstein (1952), however, while accepting that Ewing's tumour arises from the reticulo-endothelial elements in the bone marrow, regarded it as a separate entity from reticulo-sarcoma of bone.

CASE REPORTS

Case 1—Man aged thirty-one years. Laboratory assistant.

History—This man was first seen in June 1953 complaining of pain in his back for eighteen months. The pain came on after helping to move a piano. It persisted throughout 1952, but became more intense and " girdle " in character during February 1953. A radiograph taken at that time was thought to show a crush fracture of a vertebra.

On examination, he looked ill and there was spastic paralysis of the lower limbs. The level of

FIG. 1
Case 1—Initial radiographs of spine.

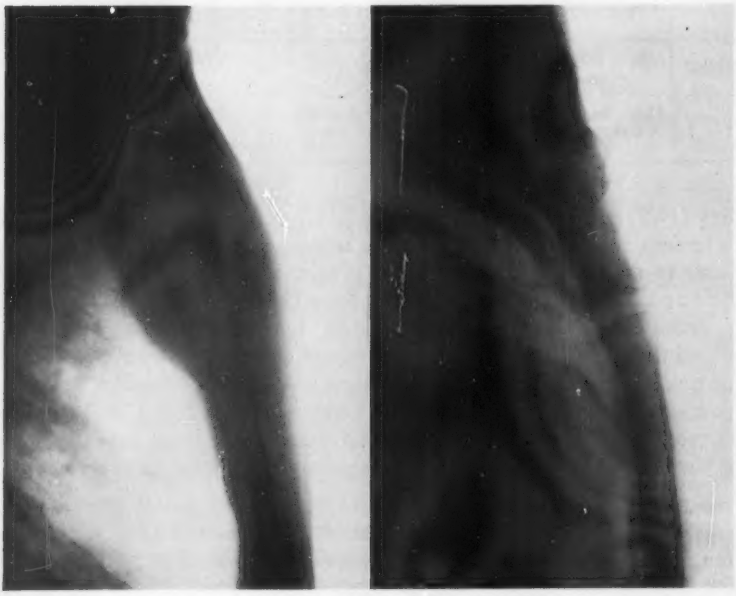

FIG. 2
Case 1—Sternum, with adjacent soft-tissue shadow seen before radiotherapy (*left*) and after radiotherapy (*right*).

anaesthesia extended up to T.11. The spinous process of this vertebra was tender, and movement at this level was limited. There was no fever. The sternum was the site of a large hard swelling in the manubrium. This was not tender.

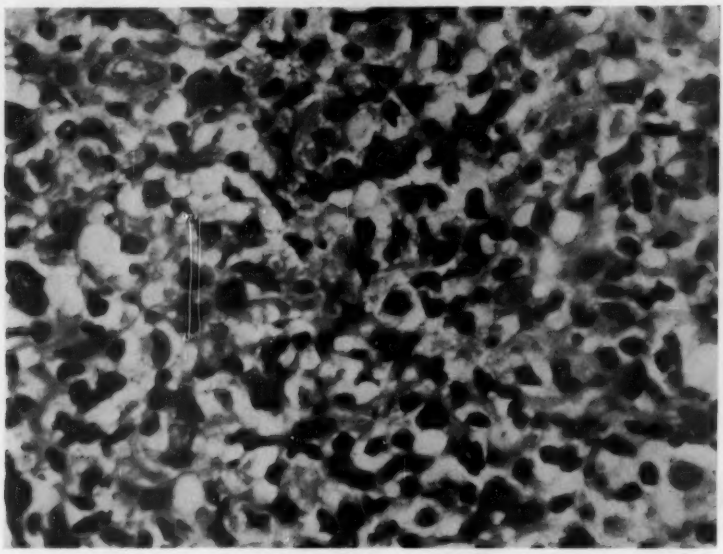

FIG. 3
Case 1—The pleomorphic pattern of the tumour is well seen. (×650.)

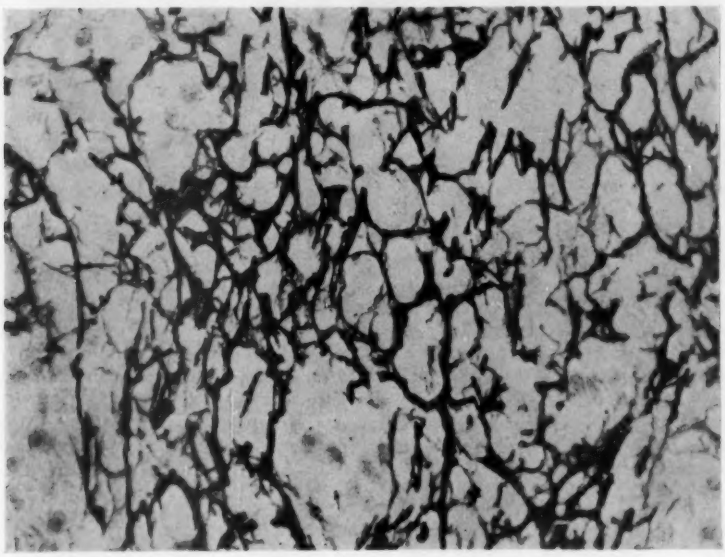

FIG. 4
Case 1—A silver impregnation to show the fairly well developed reticulin fibre network in the tumour. (×450.)

Radiographs showed wedge-collapse of the vertebral body, with condensation of bone (Fig. 1). The disc spaces were intact. For some months this was considered to be a crush fracture. The clue to the diagnosis is to be seen in the antero-posterior radiograph in which a paravertebral soft-tissue

swelling was visible. The radiograph of the sternum showed some bone destruction and a large soft-tissue swelling (Fig. 2).

Investigations—The urine contained no Bence-Jones protein. Blood examination showed: alkaline phosphatase 4·5 King-Armstrong units/100 millilitres; acid phosphatase 1·0 King-Armstrong units/100 millilitres; haemoglobin 16·3 grammes per 100 millilitres; white blood cells 5,800 per cubic millimetre.

Biopsy—The specimen consisted of several irregular fragments of firm white tissue, the largest measuring 1·0 centimetres in diameter, together with a small fragment of cancellous bone. Sections showed a somewhat pleomorphic picture (Fig. 3). The predominant cell resembled a reticulum cell but there were all gradations in size to lymphocytes. There was an occasional multinucleate cell. The nuclear pattern was variable and mitoses were infrequent. There was a rich reticulin fibre content (Fig. 4). The tumour was not unduly vascular. Conclusion: the histological picture was that of a polymorphic reticulo-sarcoma.

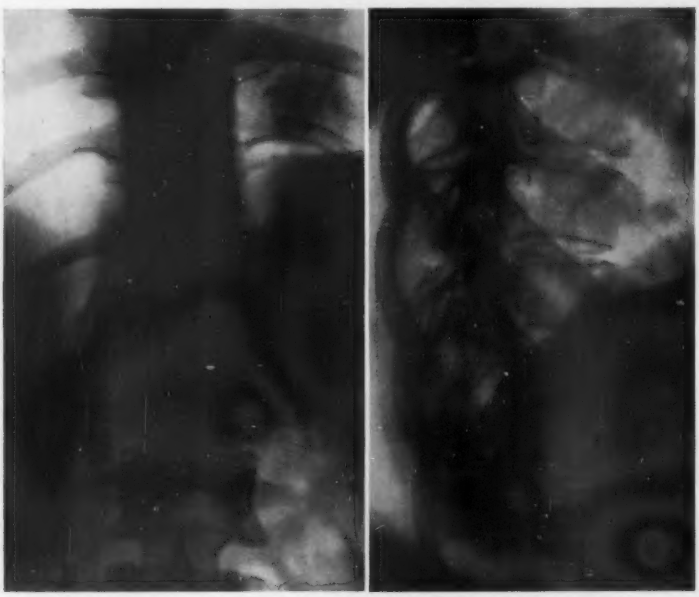

FIG. 5
Case 2—Initial radiograph of spine.

Operation (June 5, 1953)—Costo-transversectomy. A large mass of soft tumour was found in the mediastinum surrounding the vertebral body. A limited antero-lateral decompression was performed and material was removed for examination.

Post-operative radiotherapy—1,400 r in one week at 180 kilovolts to the sternum. 1,800 r in two and a half weeks at 220 kilovolts to the vertebra.

Progress—Five months after operation the patient returned to work wearing a spinal brace and walking with two sticks. Three months later he was short of breath, the abdomen was distended, and enlarged lymph nodes were present in the axillae and groins. Radiographs of the chest showed metastatic deposits in the right lung. The patient died in April 1954. Necropsy was not performed.

Case 2—Girl aged seventeen years. Shop assistant.

History—In December 1949 she fell from her bicycle and felt pain in the lower part of the back. After a week's rest she was able to return to work. In February 1950 the pain returned and she spent two weeks in bed. In March 1950 she noticed increasing weakness of her legs and difficulty with micturition.

On examination, there was a flaccid paralysis of the lower limbs and retention of urine. No local tenderness of the spine could be found.

Radiographs (Fig. 5) showed extreme collapse of the twelfth thoracic vertebra. The remains of the

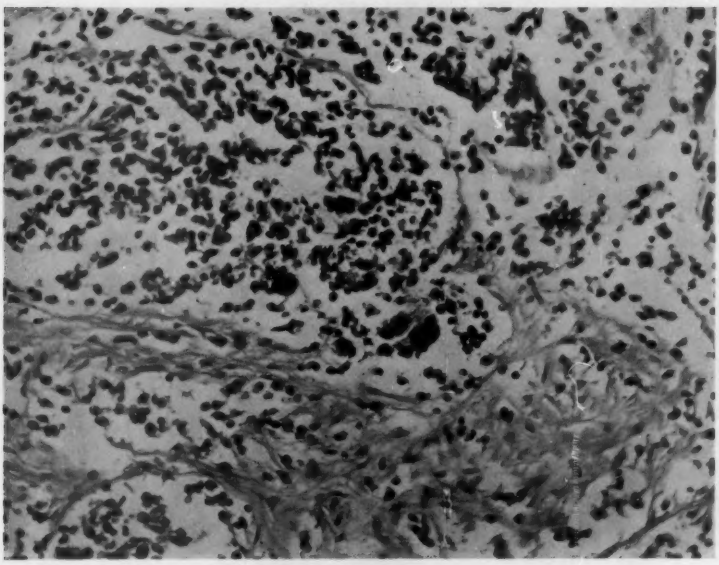

FIG. 6
Case 2—The pleomorphic cytology is well shown, several small multinucleated giant cells being present in the centre. (×170.)

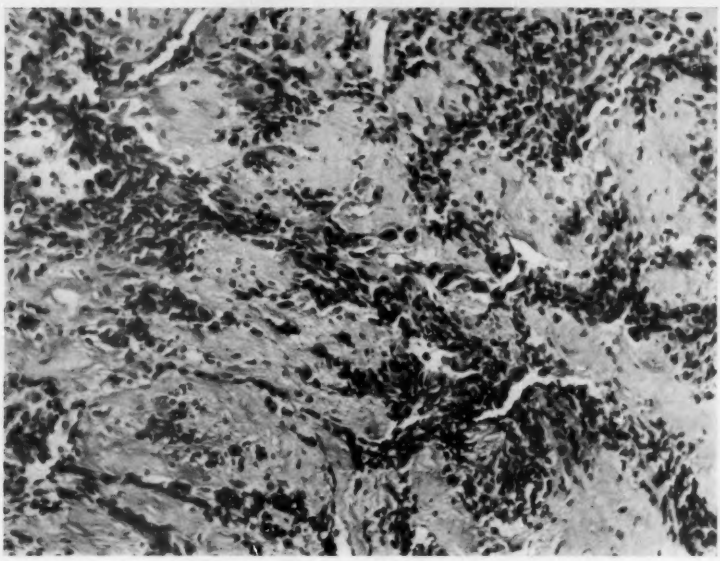

FIG. 7
Case 2—Showing the considerable fibrous stroma. (×170.)

vertebral body were seen as a dense shadow resembling Calvé's disease. In the antero-posterior film a paravertebral shadow was visible. There were deposits of growth in the tarsal bones of the left foot.
Investigations—The urine contained no Bence-Jones protein. Albumen was present, and there was infection with staphylococcus aureus and pyocyaneus. The blood showed: haemoglobin 13·3 grammes per 100 millilitres; white blood cells 13,200 per cubic millimetre; erythrocyte sedimentation rate (Westergren) 1 millimetre in 1 hour. Marrow puncture showed no neoplastic cells.
Biopsy—The specimen consisted of numerous small irregular fragments of pale brown opaque tissue in which there were a few yellow streaks. Sections showed a pleomorphic picture similar to that seen in polymorphic reticulo-sarcomas arising in soft tissue (Figs. 6 and 7). The growth had considerable fibrous stroma and was rich in reticulin fibre.
Operation (March 24, 1950)—Antero-lateral decompression was followed by little improvement of her condition.
Post-operative radiotherapy—3,200 r in two weeks at 180 kilovolts to the spine. 2,000 r in ten days at 180 kilovolts to the foot.
Progress—Two months after operation there were widespread deposits in the lungs. The patient died in June 1950.

FIG. 8

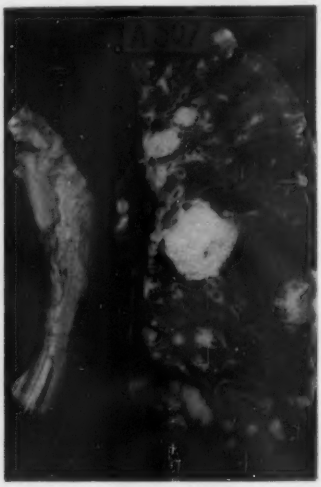

FIG. 9

Case 2. Figure 8—Complete destruction of the twelfth thoracic vertebra by firm fleshy growth. Figure 9—Numerous metastases in the lung and a large secondary deposit in a rib.

Necropsy—The striking feature was the extent of involvement of the skeleton by metastatic growth. Secondary deposits were present in all the thoracic and lumbar vertebrae and in most of the ribs. There were also extensive deposits in the head and upper part of the shaft of the left femur and in the right middle and lateral cuneiform bones. The primary tumour (Fig. 8) had completely replaced the twelfth thoracic vertebra and growth extended into the spinal theca, compressing the spinal cord. There was no mediastinal mass and no suggestion that this tumour had arisen primarily from the adjacent soft tissue. There were extensive secondary deposits in the lungs and pleurae (Fig. 9). Everywhere the growth was firm in consistency and whitish in colour. No growth was found in any groups of lymph nodes except small deposits in the tracheo-bronchial lymph nodes. Careful search was made throughout the body for any small primary carcinoma, but none could be found and the suprarenals were normal. The histological picture in all sites of growth was similar to that seen in the original biopsy of the primary growth.

Case 3—Girl aged seventeen years. Shop assistant.
History—This patient was first seen in October 1951 complaining of swelling of her left thigh. The swelling had been present for two months and had rapidly increased in size. Pain was absent except for an occasional ache.
On examination, there was a firm swelling of the lower third of the left thigh which encircled the bone

(Fig. 10). It was not tender and there was no enlargement of lymph nodes. Considerable fever was present.

Radiographs showed slight elevation of the periosteum with new bone formation (Fig. 11). This might well have been caused by infection. Six weeks later there was a large periosteal soft-tissue mass containing " sunray " spicules of new bone (Fig. 12). The radiological appearance was that of an osteogenic sarcoma.

Investigations—The urine contained no albumen or Bence-Jones protein. The blood showed: haemoglobin 10·7 grammes per 100 millilitres; white blood cells normal count; erythrocyte sedimentation rate (Westergren) 26 millimetres in 1 hour.

Biopsy—The specimen consisted of an irregular piece of soft friable tissue and a lymph node 1·0 centimetres in diameter. Section showed a very necrotic growth. In surviving areas the tumour was seen to be composed of a reticulum type of cell tending to be arranged around blood vessels (Fig. 13). The nuclei were large and hyperchromatic and mitotic figures were numerous. Little or no stroma

FIG. 10

FIG. 11

FIG. 12

Case 3. Figure 10—Photograph showing the swelling of the thigh. Figure 11—Initial radiograph. Figure 12—Six weeks later.

was present. Section of the lymph node showed no evidence of tumour. Conclusion: a syncytial reticulo-sarcoma.

Radiotherapy—2,700 r in three weeks at 220 kilovolts.

Progress—After treatment the tumour was much reduced in size and the patient gained weight. Radiographs showed that the new bone formation was circumscribed and of increased density (Fig. 14). The patient remained well for fourteen months but then complained of pain in her left thigh. On further examination in January 1953 a soft-tissue recurrence was found in the site of the biopsy scar. The histology of a further biopsy was like that of the primary lesion. The recurrence was treated by radiotherapy, 2,900 r in one week at 220 kilovolts.

Operation (February 1953)—Disarticulation at the left hip.

Pathology—A longitudinal section through the femur and soft tissue (Fig. 15) showed the bone marrow to be soft and red in the head, neck and upper part of the shaft, and soft and yellow in the mid-shaft. The marrow cavity in the lower third of the femur was replaced by dense bone. On the lateral surface of the femur in this region the periosteum was raised by a small area of soft growth containing spicules of bone. On the medial aspect of the femur there was a subperiosteal mass of growth extending

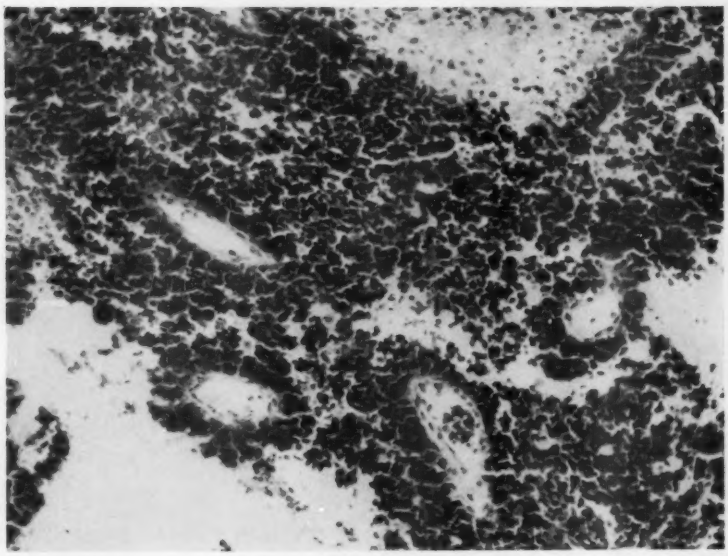

FIG. 13
Case 3—Several blood vessels surrounded by growth of a uniform cell pattern. (×170.)

FIG. 14
Case 3—Radiographs of femur after radiotherapy.

down to the medial condyle and about half-way round the circumference of the shaft. Its cut surface was white and the consistency soft. Six lymph nodes were present in the inguinal fat. Section of the marrow cavity in the lower third of the femur showed masses of new bone and no tumour tissue. Sections of the main subperiosteal mass showed a very similar picture to that seen in the biopsy but, in some sections, the cell pattern tended to be considerably more pleomorphic. There was still no reticulin fibre present. There was no suggestion of osteoid tissue or other evidence of bone formation on the part of the tumour. Sections of the lymph nodes showed no evidence of metastasis.

Progress—Three months after amputation she complained of pain in the back. Radiographs showed collapse of the eleventh thoracic vertebra. She developed swellings in the skull, frontal and occipital regions with radiological evidence of growth (osteolytic). There was cerebral compression, proptosis and papilloedema. The patient died in June 1953. Necropsy was not undertaken.

Case 4—Man aged thirty-nine. Window cleaner.

History—This patient complained of periodic attacks of pain and swelling in the upper end of the right tibia for four years. The swelling first arose while he was in

FIG. 15

FIG. 15

Case 3—Longitudinal section through femur showing the subperiosteal mass of growth partly enveloping the bone. Note the replacement of growth in the marrow cavity by new bone.

India and was described as painful " red spots." These were thought to be inflammatory and were treated with fomentations. They soon disappeared but six weeks later came back and again got better with rest and compresses. There had been no injury. Over the next two years recurrences

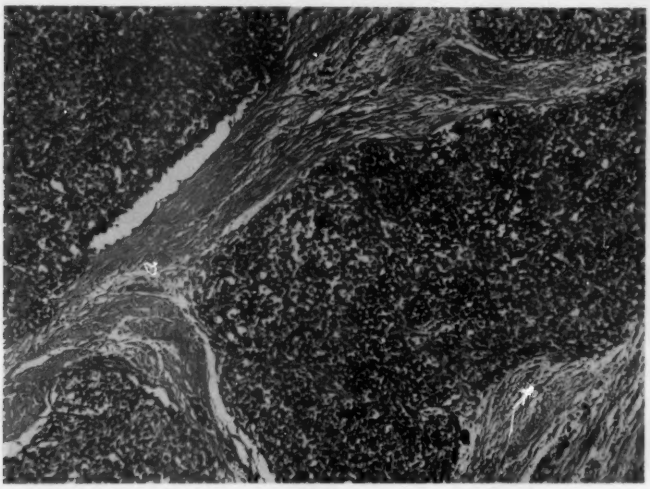

FIG. 16

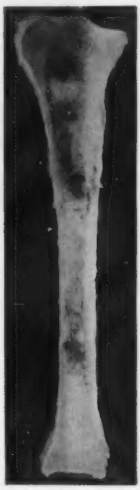

FIG. 17

Case 4. Figure 16—Masses of tumour cells separated by trabeculae of well formed collagen. (×70.)
Figure 17—Longitudinal section of tibia showing two areas of growth in the medullary cavity. The upper mass is infiltrating the cortical bone.

became more frequent, and in July 1946 he returned to the United Kingdom and entered hospital. A diagnosis of osteomyelitis was made. Treatment given was penicillin and poultices.

On examination (January 1948), there was a diffuse red swelling over the subcutaneous border of the upper third of the right tibia, measuring two and a half inches by one and a half inches. The swelling was tender, firm and lobulated. The inguinal lymph nodes were palpable and tender.

Radiographs (report only available) showed diffuse osteoporosis of the upper tibia, with periostitis of the tibia and fibula. No other radiological changes were present in the skeleton.

Investigations—The blood showed: haemoglobin 12·7 grammes per 100 millilitres; white blood cells

10,900 per cubic millimetre; Wassermann reaction negative. Biopsy of an inguinal lymph node showed " non-specific inflammation."

Biopsy of tumour—Sections of the tumour tissue removed from the centre of the bone and from the subperiosteal region showed the histological picture of a trabeculated reticulo-sarcoma (Fig. 16). The subperiosteal tumour showed no striking evidence of recent rapid growth—mitotic figures were sparse and many of the vessels in the fibrous tissue showed obliterative endarteritis. The tumour tissue from the marrow cavity, however, was much more vascular and showed numerous mitotic figures. There was practically no reticulin fibre in the tumour cell masses.

Treatment—Radiotherapy 3,200 r in two weeks at 180 kilovolts to the tibia. Two weeks later, above-knee amputation (March 1948).

Pathology—On dissection the only abnormality was a slight fusiform enlargement of the upper third of the shaft of the tibia. One small lymph node was present in the popliteal space. On sawing through the tibia longitudinally two areas of soft haemorrhagic tumour tissue were found in the medullary cavity (Fig. 17). The uppermost was 9·0 centimetres in length and corresponded to the external fusiform enlargement which was due to thickened cortical bone. The second was at the lower end

FIG. 18

Case 5—Initial radiograph showing lesion in fibula.

of the shaft and was circular, measuring 1·0 centimetres in diameter. The cortical bone overlying the upper growth was being infiltrated. Sections from the tumour mass at the upper end of the medullary cavity showed much of the growth to be destroyed by radiotherapy and replaced by fibrous tissue, but considerable areas of viable growth were still recognisable. Section from the other area of growth showed a more pleomorphic picture than that seen in the original biopsy. While there was still little stroma and no reticulin fibre the cells varied in size and shape and the nuclear pattern was much less constant. Section of the lymph node showed no abnormality.

Progress—Six years after amputation the patient was alive and well, working as a lift attendant.

Case 5—Married woman aged twenty-eight, with four children.

History—She complained of swelling of the outer side of the left lower leg of seven months duration. Size increasing, with some pain.

On examination, there was a swelling, three inches by two inches, situated in the middle third of the fibula, fixed to bone, tender, but not hot. There was no fever, and no enlargement of lymph nodes.

Radiographs showed an osteolytic lesion of the fibula (Fig. 18). In some areas there was a suggestion of expansion of the cortex. The extent of the radiological lesion should be compared with that seen in the pathological specimen (Fig. 19).

Investigations—Blood examination showed: haemoglobin 11·9 grammes per 100 millilitres; white blood cells 11,000 per cubic millimetre.
Operation (February 1947)—Excision of the mid-shaft of the fibula together with adherent muscles into which the growth was infiltrating.
Pathology—The specimen (Fig. 19) consisted of a portion of fibula measuring 13·0 centimetres. For the most of its length it was enveloped by a fusiform mass to which muscle was attached. The fibula was fractured about the centre of the specimen. On cutting the bone and tumour through longitudinally most of the bone was seen to be destroyed and replaced by a firm fleshy tumour which had invaded the periosteum and infiltrated the adjacent muscle. It extended up and down the marrow cavity to the ends of the specimen. In some areas the tumour was haemorrhagic but for the most part it was yellowish in colour. Sections showed the growth to consist of masses of cells consisting almost entirely of nucleus, there being little cytoplasm and the cell boundaries being difficult or almost impossible to define (Fig. 20). The nuclei were pale and vesiculated: mitotic figures were infrequent. The cell masses were separated by large blood sinuses, many of which had no lining apart from tumour cells (Fig. 21). There was no necrosis. The growth had practically no supporting stroma and

FIG. 19

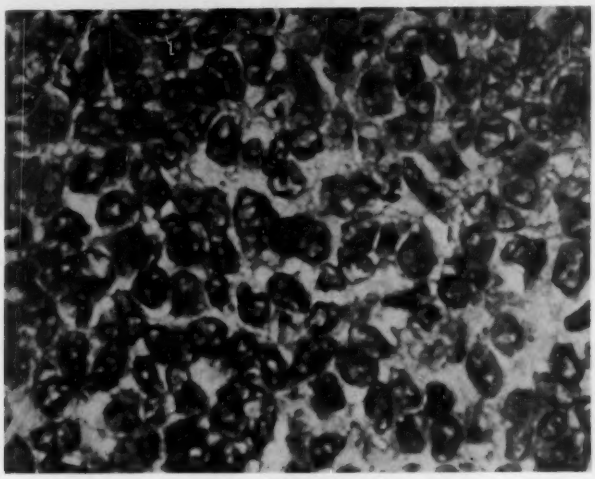

FIG. 20

Case 5. Figure 19—The fibula showing the extensive destruction of bone by growth which extends through the periosteum into the adjacent soft tissue. Figure 20—Photomicrograph showing the almost complete absence of cell boundaries and the vesicular nuclei surrounded by very little cytoplasm. (×700.)

little or no reticulin fibre was present. Conclusion: a syncytial reticulo-sarcoma arising in the bone marrow. This growth was likely to be very radiosensitive.
Post-operative radiotherapy—2,150 r in two and a half weeks at 180 kilovolts to the operation site.
Progress—When seen seven and three-quarters years after operation this patient was well. She has had another child since the operation.

Case 6—Married woman aged thirty-eight.
History—This patient complained of a lump in the sternum increasing in size for two years. At first it was painful, but latterly not so. In July 1952 a biopsy of the sternal swelling was carried out, and the histological report suggested " chronic infection."
On examination (December 1952), there was a hard tumour projecting from the sternum, and measuring two inches by two inches. A retrosternal swelling could be felt in the suprasternal notch.
Investigations—Blood examination showed: haemoglobin 12·1 grammes per 100 millilitres; white blood cells 7,000 per cubic millimetre; erythrocyte sedimentation rate (Westergren) 6 millimetres in 1 hour. Wassermann reaction negative.
Radiographs (Fig. 22) showed an osteolytic lesion in the sternum with a large soft-tissue mass (compare with Case 1, Figure 2). Intravenous pyelography showed normal kidneys. The chest radiographs were normal.
Biopsy—Sections showed tumour masses separated by irregular trabeculae of collagen. The tumour

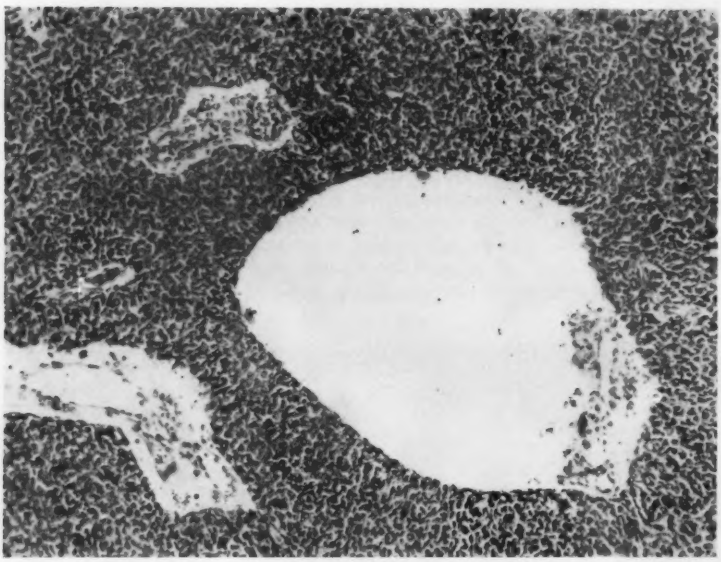

FIG. 21
Case 5—Showing a large blood space entirely surrounded by tumour cells. (×110.)

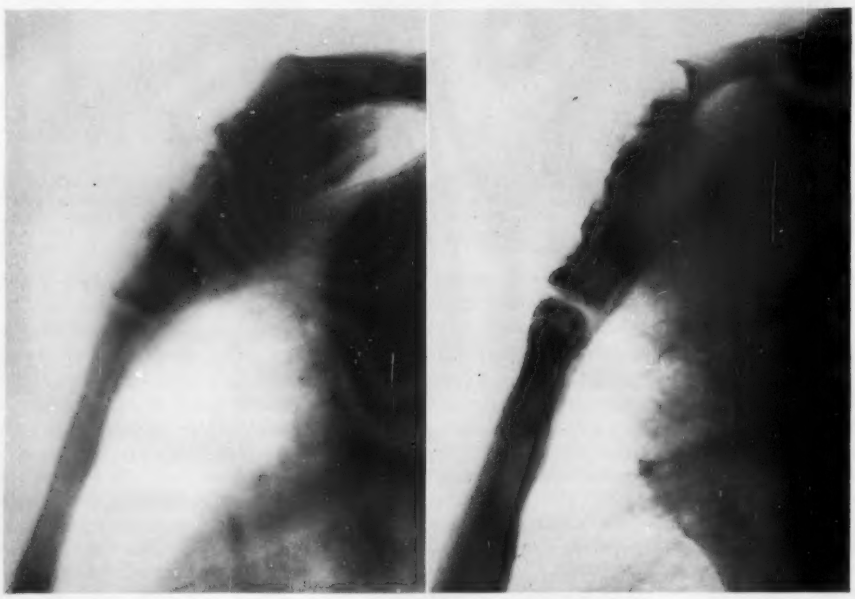

FIG. 22
Case 6—Radiographs of sternum before radiotherapy (*left*) and after radiotherapy (*right*).

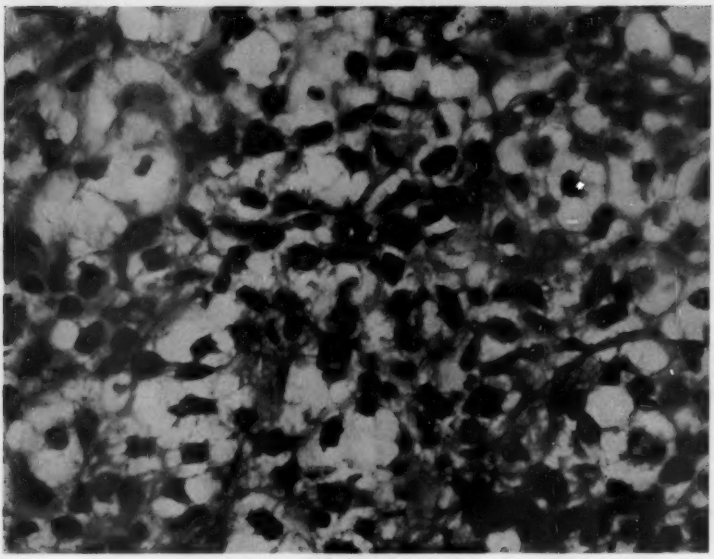

FIG. 23
Case 6—Section showing a pleomorphic structure like that seen in Case 1.

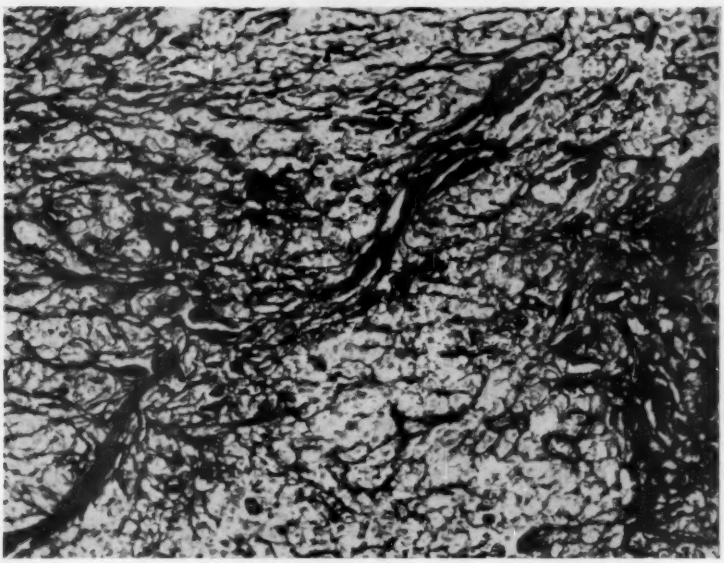

FIG. 24
Case 6—A silver impregnation showing the rich reticulin content of the tumour. (×170.)

FIG. 25
Case 7—Radiograph showing lesion in the acromion process.

showed a very pleomorphic picture (Fig. 23) which was very similar to that seen in a polymorphic reticulo-sarcoma arising in lymphoid tissue. It was very rich in reticulin fibre (Fig. 24). The histology was almost identical to that seen in Case 2.
Treatment—Radiotherapy only. 4,000 r in forty-two days to the sternum.
Progress—When seen two years after treatment the patient was well, and the swelling had disappeared.

TABLE II
SUMMARY OF MAIN CLINICAL FEATURES IN SEVEN CASES OF RETICULO-SARCOMA OF BONE

Case	1	2	3	4	5	6	7
Site	Spine-sternum	Spine	Femur	Tibia	Fibula	Sternum	Acromion
Age	31	17	17	39	28	38	22
Sex	Male	Female	Female	Male	Female	Female	Female
Symptom	Pain	Pain	Swelling	Pain and swelling	Swelling	Swelling	Pain
Duration	18 months	2 months	2 months	48 months	7 months	24 months	10 months
Treatment	Radiotherapy	Radiotherapy	Radiotherapy. Amputation	Radiotherapy. Amputation	Excision. Radiotherapy	Radiotherapy	Excision. Radiotherapy
Follow-up	*Died* 28 months	*Died* 7 months	*Died* 22 months	*Alive* 10 years	*Alive* 8 years	*Alive* 3½ years	*Alive* 3 years
Time since first symptom	28 months	7 months	22 months	11 years	8½ years	3½ years	3 years
Cause of death	Deposits in lungs	Multiple deposits (autopsy)	Multiple deposits. Skull. Spine	—	—	—	—

Case 7—Married woman aged twenty-two.
History—In May 1953 she complained of pain in the right shoulder of gradual onset for ten months. Pain was severe for the first three weeks, but afterwards only intermittent.
On examination, the patient was four months pregnant. The acromion process was tender but there was no swelling. Movements of the shoulder were slightly limited and there was a little wasting of the deltoid and spinati muscles.
Radiographs showed mottling of the right acromion process (Fig. 25). No soft-tissue swelling was visible. The provisional diagnosis was " tuberculosis."
Investigations—The urine contained no albumen or Bence-Jones protein. Blood examination showed: haemoglobin 14·2 grammes per 100 millilitres; white blood cells 11,200 per cubic millimetre; erythrocyte sedimentation rate (Westergren) 15 millimetres in 1 hour.
Operation (June 1953)—Excision of acromion process.
Pathology—The terminal part of the acromion was expanded and a large amount of thickened fatty tissue was adherent to its surface. On cutting the acromion longitudinally it was seen that much of it was replaced by firm fleshy tumour tissue showing no necrosis. The growth did not appear to extend through the covering periosteum. Sections showed the histological picture of a polymorphic reticulo-sarcoma. There was a rich reticulin fibre network between the cells of the growth.
Post-operative radiotherapy—1,300 r in two weeks at 180 kilovolts.
Progress—The patient was well one year after treatment.

CLINICAL FEATURES

Consideration of these seven cases emphasises certain points made by previous authors. The site of the tumours is shown in Figure 26. There were three tumours in the lower limbs, two in the spine, one in the sternum, and one in the acromion process. The latter is an unusual position. Of the patients with vertebral tumours, one had developed a sternal swelling before admission to hospital, but this was not until eighteen months after his first symptom in the spine. The insidious onset of the disease is well illustrated by these two spinal cases, in which backache has been present for several months, yet only when paraplegia became obvious did these patients seek hospital care. The first symptom was swelling in three cases and pain in three. One patient complained of both. The tumour of the femur (Case 3) was the only one to show a daily variation in size, which is mentioned in the literature as a feature of reticulo-sarcoma of bone.

The duration of symptoms varied between two and forty-eight months, the average being sixteen months. In the early stages the similarity to chronic bone infection was striking. This is well illustrated in the swelling of the tibia (Case 4) which had been treated as osteomyelitis for four years and in the sternal swelling (Case 6), the first biopsy of which was reported as showing " chronic infection."

Fever was present as an early symptom in only one patient (Case 3). In the late stages of the disease it was a result of urinary infection, when paraplegia had completed the picture (Cases 1 and 2). The good general health of these patients in relation to the size of the tumour is mentioned in the literature as a feature of reticulo-sarcoma. This was found to be true, and only when general dissemination had taken place did the patient begin to look ill or to lose weight. The same can be said in regard to lymph node involvement by the growth, as this was found in only one instance as a terminal event (Case 1).

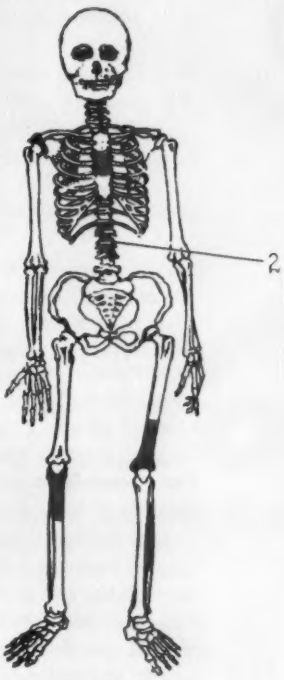

FIG. 26
Site of tumour in the seven cases of this series.

RADIOLOGICAL FEATURES

Four facets of the radiological diagnosis will be considered: 1) The diagnosis of Ewing's tumour; 2) the diagnosis of reticulo-sarcoma; 3) the radiological appearances in the present series; and 4) how far can these tumours be distinguished by radiological means?

The diagnosis of Ewing's tumour—The appearances described in Ewing's tumour are extremely variable. It " can be confused with any lesion of bone from osteomyelitis to osteogenic sarcoma " (McCormack *et al.* 1952). Lysis, mottling, sclerosis, cyst formation and periosteal reaction are described. These authors were able to make a diagnosis in only twelve out of eighty cases on a radiological basis. The so-called characteristic features are the situation of

FIG. 27
" Lamination " in an osteogenic sarcoma.

FIG. 28
" Lamination " in staphylococcal osteomyelitis.

the tumour in the shaft of a long bone, its extent, and the periosteal reaction. The well known laminated or " onion-peel " appearance of reactive bone is by no means a constant feature. When " sunray " spicules are present they suggest an osteogenic sarcoma. Expansion of the shaft is more commonly seen in Ewing's tumour (Coley 1948).

The diagnosis of reticulo-sarcoma—Typical radiological appearances of primary reticulo-sarcoma of bone are described in the literature. It is suggested that a diagnosis can be made upon radiological grounds alone in many cases (Sherman and Snyder 1947). The tumour is situated near the end of a long bone, but tends to extend towards the shaft. It originates in the medulla and is essentially osteolytic. The cortex is destroyed, but there is no expansion. Periosteal reaction is not a constant feature, and in fact some authors stress the absence of reactive new bone under the periosteum (Valls *et al.* 1952). A soft-tissue shadow is frequently seen in the radiograph. This contains no tumour pattern and is never calcified (Sherman and Snyder 1947), in contrast to the soft-tissue tumour shadow in osteogenic sarcoma, in which calcification is common.

The response to irradiation is characteristic. The soft-tissue shadow first disappears and this is followed by re-formation of the cortex. The medullary bone is repaired and assumes a density somewhat greater than that of normal bone.

Radiological appearances in the present series—In the two vertebral tumours (Cases 1 and 2) it is difficult to describe any special features. In both, collapse of the bodies had taken place before films were taken and the texture was that of condensed bone. A paravertebral soft-tissue shadow, together with collapsed vertebral body and intact intervertebral discs, suggested the diagnosis of a malignant tumour. The true nature of this tumour was not determined until a biopsy had been done.

The tumour of the femur (Case 3) is perhaps the most instructive. The early films showed the presence of periosteal new bone (Fig. 11) without cortical erosion. This might well have been of inflammatory origin. Six weeks later this new bone was still present above the medial femoral condyle, but there was now a large soft-tissue tumour which was ossifying with "sunray" bone spicules. These films suggested an osteogenic sarcoma (Fig. 12). The tumour proved to be radiosensitive and after treatment became circumscribed. The films should be compared with Figure 27, which shows the typical appearance of a Ewing's tumour with regard to its site, and lamination of the periosteum, but in which the histological diagnosis was that of an osteogenic sarcoma. Again, Figure 28 is a good example of laminated periostitis in a case of staphylococcal osteomyelitis.

The tumour of the fibula (Case 5) showed an osteolytic lesion with a suggestion of cortical expansion (Fig. 18). The cortex was destroyed in places but there was little reactive new bone. This film should be compared with the photograph of the specimen (Fig. 19). The radiograph does not indicate the true extent of the tumour, which occupies considerably more of the fibular shaft. On removal at operation the bone was divided through tumour tissue, yet the patient is alive more than eight years later. The radiological appearance was that of a Ewing's tumour.

The tumour of the sternum (Case 6) was an osteolytic bone lesion with destruction of the cortex, but no bone expansion. A soft-tissue mass was present (Fig. 22). The picture is almost identical with that seen in the secondary tumour of the sternum in Case 1 (Fig. 2). After radiotherapy the soft-tissue mass became smaller and there was reactive new bone in the sternum. The appearance is that of a reticulo-sarcoma.

The tumour of the acromion process showed mottled sclerosis and rarefaction. There was no periostitis and no soft-tissue mass (Fig. 25).

How far can these tumours be distinguished by radiological means?—Although they form too small a series from which to draw any sweeping conclusions, these seven cases illustrate the diversity of radiological changes, which do not conform to any one type. We are left with the impression that it is not possible to distinguish between a Ewing's tumour and a reticulo-sarcoma on radiological grounds.

Ancillary methods of investigation, which included erythrocyte sedimentation rates, blood picture and search for Bence-Jones protein in the urine, proved to be of no help in diagnosis. This must be based upon an interpretation of the history, clinical and radiological signs, and on the histological findings, the latter being of particular importance.

The differential diagnosis must include chronic osteomyelitis and periostitis, osteogenic sarcoma, secondary carcinoma, multiple myeloma, primary lymphosarcoma with bone involvement, and eosinophilic granuloma. The question of differential diagnosis between Ewing's tumour and reticulo-sarcoma will be discussed later.

PATHOLOGY

In our material it has not been possible to separate the tumours into two clear-cut groups on the clinical or radiological appearances, and the gross pathology and histological picture

also did not enable this to be done. Figure 26 shows the sites of the tumours in the seven cases. Three were in long bones (right tibia, lower third of femur and mid-shaft of fibula) and four were in short bones (two in vertebrae, right acromion and sternum).

The gross appearance of the tumours occurring in long bones did not differ remarkably from those seen in other primary osteolytic tumours. They appeared to arise either in the mid-shaft or in the metaphysis, and extended through the cortex with a variable degree of bone destruction, lifting up the periosteum and forming a subperiosteal mass which enveloped the bone (Figs. 15, 17 and 19). Although the tumours themselves were not osteogenic the periosteum might be stimulated to form bone, either in layers to produce the " onion-peel " appearance, or in a radial manner to produce the typical " sunray " appearance of a periosteal osteogenic sarcoma (Fig. 12). The growth tended to spread up and down the marrow cavity to a much greater extent than was revealed by radiology. Whilst the periosteum contained the growth for a very considerable time it might be infiltrated eventually and the surrounding soft tissues involved (Case 3, Fig. 15). The growths were reddish in colour and usually not unduly soft or necrotic unless there had been intensive recent x-ray therapy before amputation. In Case 3 (Fig. 15) the growth, present in the medullary cavity, had been almost completely replaced by new bone, but this was following intensive x-ray therapy.

When small bones such as vertebrae were involved the tumour rapidly destroyed the bone, which collapsed. The adjacent discs were not involved, but growth extended into the surrounding tissues to a varying degree to form a soft-tissue mass. When the growth arose in the acromion and in the sternum there was, however, no evidence of extension through the periosteum into the adjacent tissues. In the only case to come to necropsy (Case 2) the growth started in the twelfth thoracic vertebra. Despite radiotherapy, death took place three months from the time the diagnosis was made. At necropsy the twelfth thoracic vertebra was completely replaced by growth and there were extensive metastases in other parts of the skeleton and in the lungs. There was nothing to suggest a primary origin in lymph nodes and the suprarenals were normal. Whereas clinically this case behaved as a Ewing's tumour, histologically it would not have been accepted as such because its variable histological structure and rich reticulin fibre content classes it as a reticulo-sarcoma (Figs. 6 and 7).

In the seven cases studied the histological structure was variable. In some the emphasis was on solid masses of tumour cells separated by trabeculae of fibrous tissue and little or no reticulin between the tumour cells (Fig. 16), whereas in others the picture was more pleomorphic with considerable collagen and reticulin fibre present (Figs. 3 and 4). As in most malignant tumours the apparent constancy of the histological picture in any one case varied with the number of sections examined from different areas of the growth.

In those growths in which much of the tumour had a fairly uniform structure and conformed most closely to that described as typical of Ewing's tumour the masses of tumour cells showed no cell boundaries and their nuclei were large and vesiculated and had an average diameter of 8 microns. They were surrounded by only a small quantity of cytoplasm. Nucleoli were uncommon and mitoses not very frequent. There was little or no reticulin fibre in these masses of tumour cells. In more pleomorphic areas and in those growths which throughout had a much more variable structure there was a rich reticulin stroma. The tumour cells varied in size and shape from occasional multinucleate cells to cells the size and shape of lymphocytes, but the predominant cell was considerably larger than a lymphocyte with a round or oval nucleus. The cell boundary was surrounded by a varying quantity of cytoplasm, which was usually basophilic but occasionally acidophilic. Nucleoli were infrequent, but mitoses were quite numerous. In none of these cases were any structures resembling rosettes seen. In all seven cases the tumours had a rich blood supply. In one case (Case 5) there were many blood spaces lined by tumour cells (Fig. 21), yet this patient is alive and well more than eight years from the time the diagnosis was made. In no case was there any evidence of bone formation on the part of the tumour.

DISCUSSION

The impression gathered from the literature that the separation of Ewing's tumour and reticulo-sarcoma was an artificial one has been confirmed by a study of this small series of cases.

The age of our patients ranged from seventeen to thirty-nine years; five were females and two were males. There was no striking difference in the mode of onset of the disease or in the duration of symptoms from those described in the literature. Three tumours occurred in long bones and four in other parts of the skeleton. This series is, however, too small to draw any conclusions from these facts. In only one case was fever present early in the disease and in all the blood counts and urine were normal.

Radiologically these seven cases illustrate a diversity of changes which do not conform to any one type. The impression gained is that it is not possible to distinguish between a Ewing's tumour and a reticulo-sarcoma of bone on radiological grounds. When radiological and histological changes are considered together it will be seen that this distinction is even more difficult.

The gross appearance of the tumours was variable and might even resemble that of an osteogenic sarcoma. This was particularly so in long bones. The histological picture was inconstant and, in this respect, resembled that seen in reticulo-sarcomas arising in lymphoid tissue. In three cases the histology, on the whole, was that of a Ewing's tumour as described in the literature, yet two of these patients are alive and well eight and eleven years after their first symptoms. In four cases in which pleomorphism was marked and reticulin fibre content well developed two patients died rapidly.

The criticism could be made that all these cases were reticulo-sarcomas of lymphoid tissue with early manifestations in the skeleton. The clinical picture is against this because no lymph node enlargement was found except in one case as a terminal event when there were general metastases. Also, in no case was a mediastinal or retroperitoneal mass demonstrated at any time. Besides, involvement of the skeleton in reticulo-sarcomas arising in soft tissue is uncommon. Craver and Copeland (1934) investigated the incidence of involvement of bone in soft-tissue lymphosarcomas either directly or by haematogenous spread. In 164 cases only seventeen showed lesions in bone. Of these, all except one had lymph node involvement before there was evidence of a bone tumour.

Similarly the clinical course and pathological picture rules out the possibility that these cases were unusual examples of carcinomatosis or secondary neuroblastomas. While it is true that there was no necropsy on two of them, one patient was aged seventeen and the other aged thirty-one, and reference to their case histories and pathology will show that these assumptions are most unlikely.

We consider therefore that a case has been made out for the recognition of primary reticulo-sarcoma of bone as a clinical and pathological entity. We have found no evidence to support the conception that this tumour is an entity separate from Ewing's tumour. We believe that they are one and the same thing.

Reticulo-sarcoma of bone is not a common condition, only seven examples of it having been seen in eight years. Its recognition, however, is important because the prognosis may be good, five out of seven patients being alive and well, one for eight and another for eleven years. Two patients presented, on the whole, the accepted histological picture of a Ewing's tumour, yet these have survived the longest. Two patients showed histologically a pleomorphic picture with a rich reticulin fibre content, yet they died rapidly.

The tumours are radiosensitive, and the best treatment is probably a combination of radiotherapy with excision or amputation when this is possible.

SUMMARY

1. Seven new cases of primary reticulo-sarcoma of bone are recorded, including one in which a full necropsy was carried out.

2. The literature of Ewing's tumour and reticulo-sarcoma of bone has been examined and the conclusion reached that the separation of these two tumours is an artificial one.
3. The investigation of our cases confirms this observation.

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HISTIOCYTIC GRANULOMATOSIS

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The rarity of this somewhat obscure condition, and certain interesting features to be discussed in this case, seem to warrant its publication, even although it is a solitary case.

The title, suggested in 1941 by Farber, and modified subsequently by Weinmann and Sicher (1955) is favoured because the permanent and constant pathological feature is the presence of numerous histiocytes and granulomatous tissue. These cells, in the case under consideration, have undergone a transformation from the early stage where there were numerous eosinophil cells simulating an eosinophilic granuloma, to a later stage in which are seen numerous " foam cells " typical of a lipoid granulomatosis. This interesting change has been followed through a period of six years, and from this respect is unique.

Since Fraser's review (1935) much thought has been given to the disease with its varied expressions or manifestations and its terminology. Recent reviews by Jaffe and Lichtenstein (1944), Snapper (1949), Hill (1949), Fairbank (1951), and Lichtenstein (1953), describe the origin of the varying nomenclature, such as Hand-Schüller-Christian disease, or lipoid granulomatosis, the latter name being accredited to Greig by Fraser (1935), solitary granuloma of bone (Otani and Ehrlich 1940), non-lipoid histocytosis or Letterer-Siwe disease (Abt and Denenholz 1936), eosinophilic granuloma of bone (Mallory 1942, Jaffe and Lichtenstein 1944), non-specific granuloma of bone (Hill 1949), and, most recently, histocytosis X (Lichtenstein 1953). But a marked and important advance was made by Farber's observation in 1941 that " anatomically the underlying lesion in eosinophilic granuloma is related to the lesion of Schüller-Christian disease and that of Letterer-Siwe disease." This was further expounded by Green and Farber (1942) and in great detail by Jaffe and Lichtenstein, who described these three conditions as " different clinico-anatomic expressions of the same basic disorder." There has been much literature in America and in Europe, the main contribution from the latter being from Engelbreth-Holm *et al.* (1944), describing how eosinophilic granuloma, Letterer-Siwe disease and Schüller-Christian disease were all related phases of the same condition. A more recent and critical review of this nature was presented by Lichtenstein (1953), who has attempted to clarify and to integrate these clinical expressions under the heading of histiocytosis X.

With this case, there is presented pathological evidence to support Green and Farber's original concept of the interrelationship between the conditions of eosinophilic granuloma and lipoid granulomatosis, as well as observations on pathogenesis and treatment of this condition.

CASE REPORT

In July 1949 a married woman aged thirty-one years was admitted to hospital complaining of pain in the left hip, thigh and knee regions. This was aching in quality and there had been a gradual loss of weight in the last six months.

On examination it was noted that there was a slight degree of exophthalmos present. There was local tenderness and there was limitation of all movements of the left hip.

Haematological investigations were normal except for the erythrocyte sedimentation rate, which was 50 millimetres in the first hour, and there was a hypochromic anaemia. There was no abnormality of the chemistry of the blood.

Radiographs showed " cystic " areas in the skull, pelvis, femora, scapulae and right humerus.

Biopsy—A biopsy was carried out and the pathological report was: " The specimen consists of vascular granulation tissue which is heavily infiltrated with polymorphs, lymphocytes and histiocytes. A few giant cells of foreign-body type are seen. Many of the polymorphs are

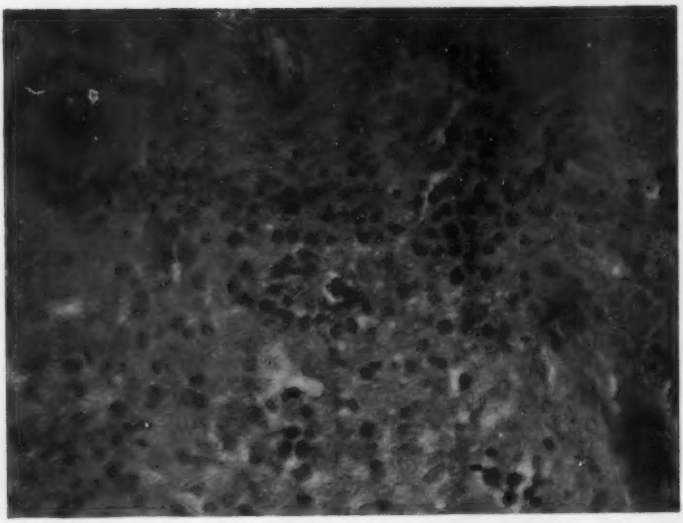

FIG. 1

Photomicrograph of section from the left ilium showing vascular granulation tissue, heavily invaded by eosinophil polymorphs, lymphocytes and histiocytes. (Haematoxylin and eosin, ×400.)

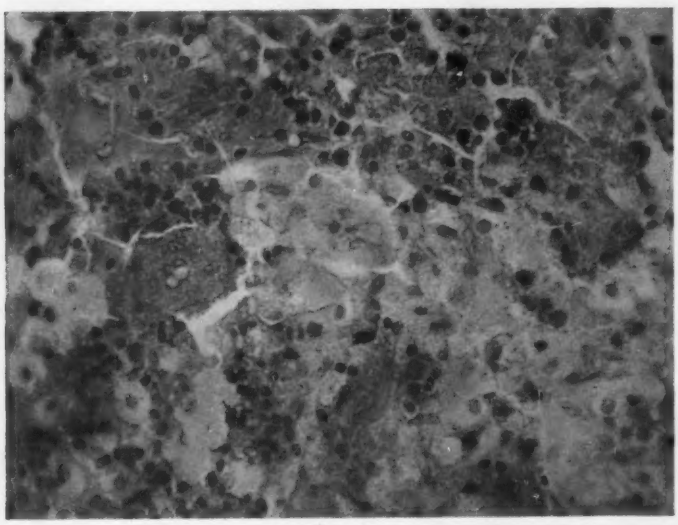

FIG. 2

Photomicrograph from same biopsy, showing area of necrosis with surrounding "foamy" cells and eosinophil polymorphs. (Haematoxylin and eosin, ×400.)

eosinophils (Fig. 1) which are widely distributed, and a large proportion of histiocytes which are 'foamy' cells (Fig. 2) are often conspicuously grouped. The 'foamy' cells and histiocytes (Fig. 3), however, dominate the picture and are often massed in sheets apart from the necrotic areas.

"The features are of a histiocytic granulomatous condition with eosinophilic and foamy characters—the latter feature would appear to be secondary, at least in part, to the necrosis. In conclusion, the condition is classified as a histiocytic granuloma."

Treatment—Deep x-ray therapy was given, with improvement in the symptoms of pain after an initial aggravation. The pain recurred however and continued throughout 1949. The weight continued to fall.

In November 1950, on falling from a bus, she sustained a transverse fracture of the right femoral neck. A radiograph showed this to be through another area of diseased bone. She was immobilised in a plaster hip spica for six months without any radiological evidence

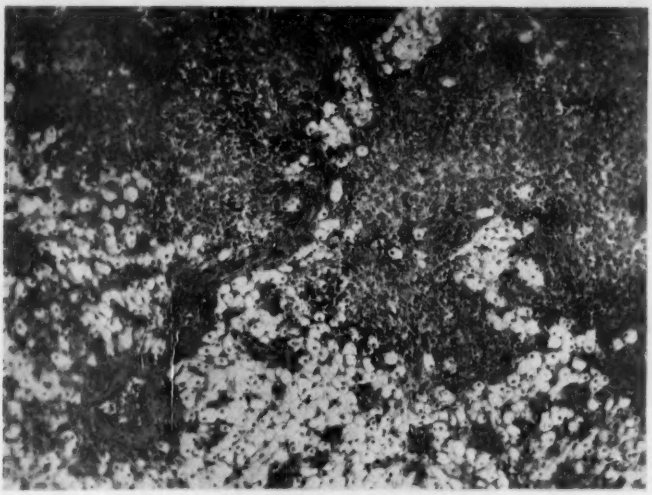

FIG. 3
Photomicrograph of same area showing sheets of histiocytes and "foamy" cells. (×100.)

of bone healing. In spite of this she was allowed up in a walking caliper and for six months she was ambulant.

In 1951 the pain became more acute in the right hip region, and she sustained a pathological fracture—intertrochanteric in type—in the left femur, with severe pain. On reaching for the nurse's arm shortly afterwards she experienced another episode of severe pain in the right upper arm, which proved to be the result of another pathological fracture—oblique in nature—through the midshaft area of the right humerus.

This patient was then placed into a plaster posterior shell with a right-angled arm support for the fractured humerus, and in this she was nursed both in hospital and at home during the next four years.

INVESTIGATIONS—1951-1955

Radiological examinations

Skull—There are translucent areas in the frontal, parietal and occipital regions (Fig. 4), varying in shape and size, but clear-cut and extending across suture lines. There is no surrounding osteosclerosis. The margins are irregular and appear to be "geographical" in

FIG. 4
Radiograph of the skull with translucent areas in the frontal, parietal and occipital regions.

FIG. 5
Radiograph of the pelvis and upper femora showing the diseased areas, with the pathological fractures in the femora.

outline. Neither the base of the skull nor the mandible is involved. These appearances have remained unchanged during these years.

Pelvis and upper femora—Numerous but typical translucent areas, without surrounding osteosclerosis, are seen in ilium, ischium and one small area in the pubis (Fig. 5). The cyst-like appearance is marked, particularly in the left ilium. The disease process is seen in both upper femora, with pathological fractures—transcervical and intertrochanteric on the right side, and transtrochanteric with avulsion of the lesser trochanter on the left side.

In Figure 6 there is seen the post-operative appearance of the hip regions with little attempt at regeneration of bone.

Shafts of femora—Pathological fractures are seen in the upper parts, through translucent, cyst-like areas. But in the shafts distal to these areas there is marked loss of thickness of cortex with an abnormal, striated appearance overlying the medullary cavity (Fig. 7).

FIG. 6
Radiograph six months after operation showing little bone regeneration.

Shoulder girdles (Figs. 8 and 9)—The condition is seen in the acromion and coracoid processes as well as the neck and body of the scapulae. Both upper humeri are diseased and in one area there is destruction and " ballooning " outwards of the cortex on the medial side without any periosteal reaction.

Humerus—Each is involved in the upper half only. The pathological and spiral fracture through the midshaft of the right humerus is seen before (Fig. 10) and after immobilisation, and is incompletely united two years later (Fig. 11). The diseased area persists as a cyst-like space.

Chest—A generalised, diffuse infiltration or fine miliary change in the lung pattern of both lung fields is noted (Fig. 12).

Pathological examinations—Biopsy material from the greater trochanter of the left femur was obtained by open operation in December 1951. " The specimen is of soft consistence and is composed in the main of a tissue made up of accumulation of vacuolated histiocytes

supported by fibroblastic granulation tissue which, in some areas, dominate the picture (Fig. 13). The histiocytes present a foamy appearance (Fig. 14), as a consequence of lipoid vacuolation. In regions where the granulation architecture predominates, leucocytic infiltration is often heavy, but the leucocytes exhibit degenerative changes and only a few can be recognised as eosinophil polymorphs or other specific varieties of cells. Irregularly scattered through the granulomatous background there are multinucleated giant cells.

FIG. 7
Radiographs of the upper femora to show the pathological fractures and the striated appearance down the shaft of each bone.

" In conclusion, the tissue from the femur is showing the lipoid storage phase of a lipoidosis, namely Hand-Schüller-Christian's disease."

Biopsy material from the greater trochanter of the left femur was obtained by open operation in February 1952.

" The tissue is largely composed of close-packed histiocytes with finely vacuolated cytoplasm. These cells are arranged in clusters supported by a delicate framework composed of fibroblasts and narrow capillaries (Fig. 15) but in all fields the histiocytic component of the tissue predominates. There is both focal and diffuse infiltration by small round cells only a few of

FIG. 8

FIG. 9

There is seen involvement of the scapulae as well as both upper humeri. On the medial side of the left humerus (Fig. 9) there is a cyst-like space with " ballooning outwards " and destruction of the cortex.

FIG. 10

FIG. 11

Shows the pathological fracture through the midshaft area of the right humerus and the amount of repair after two years.

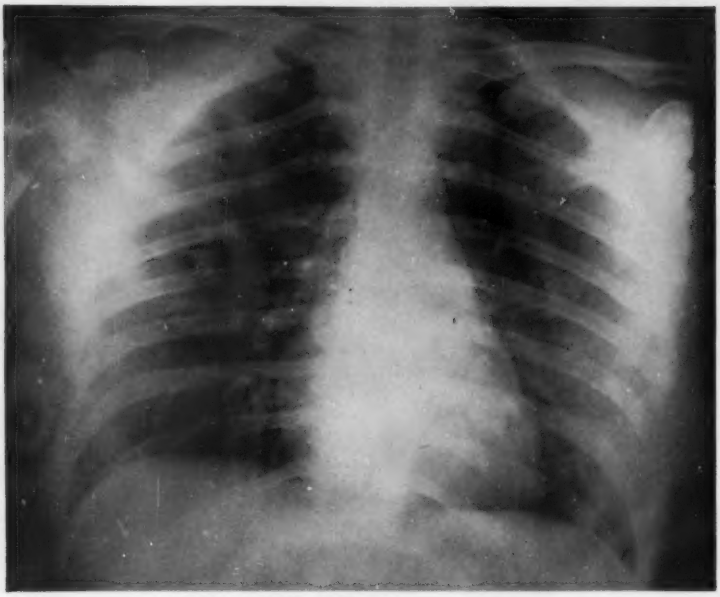

FIG. 12
Radiograph of the chest showing generalised and diffuse infiltration of both lung fields.

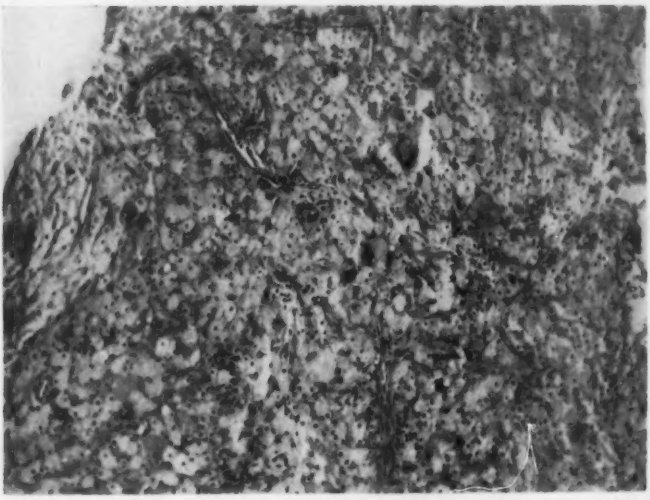

FIG. 13
Photomicrograph of a section from the left greater trochanter containing numerous vacuolated histiocytes, with some fibroblastic granulation tissue. (×100.)

which are recognisable and a scattering of giant cells. No significant difference can be detected between the tissue comprising the present biopsy and that of the previous biopsy from the same subject."

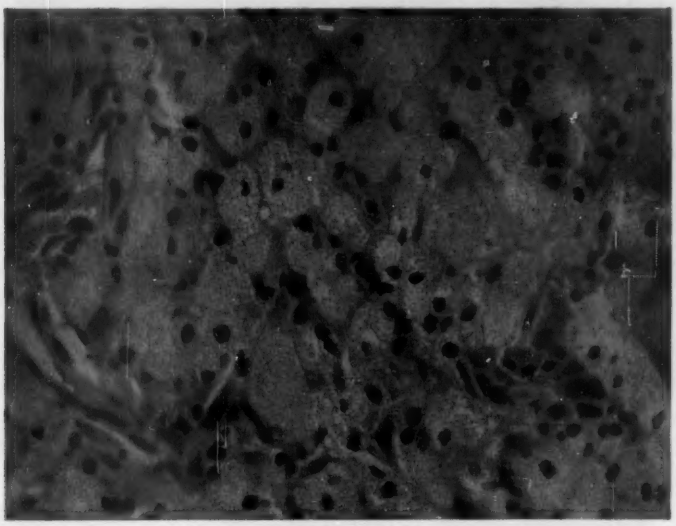

FIG. 14

High power photomicrograph of same area to show the foamy appearance of the histiocytes. (Haematoxylin and eosin, ×400.)

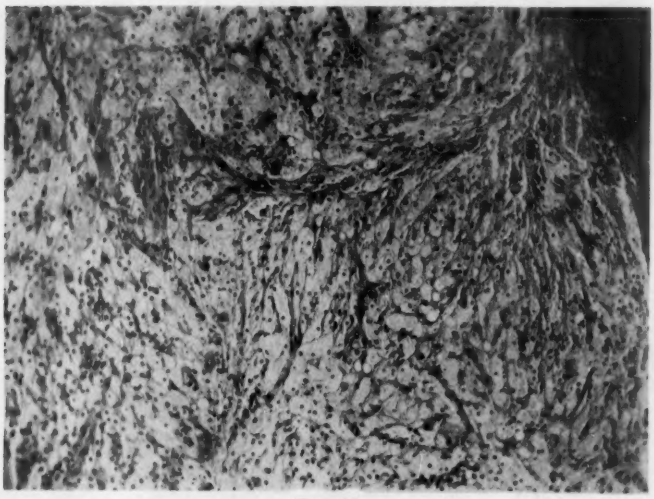

FIG. 15

Photomicrograph from the same area, but following cortisone therapy, which shows the main " histiocytic " picture to be unchanged. (×100.)

Biospy material from the upper end of the left femur was removed during radical saucerisation and a bone-grafting operation in November 1954.

" The specimen consists of a portion of bone, partly composed of thickened bony trabeculae showing some irregularity of the cement lines and very occasional lacunar erosion

(Fig. 16). Some osteoblastic activity is apparent at the margins of several of the bone trabeculae. In general the marrow is abnormal showing infiltration of foamy phagocytes (Fig. 17),

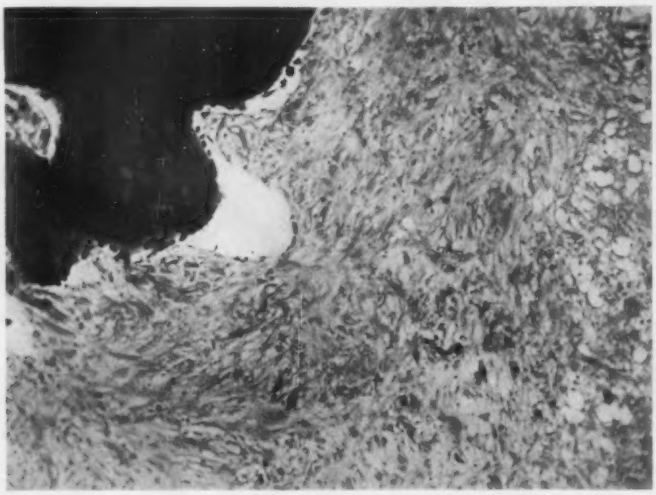

FIG. 16

Photomicrograph from the fracture area through the left femoral neck region, showing thickened bony trabeculae and some osteoblastic activity, but infiltration by " foamy " phagocytes is still marked as seen in Figure 17. (×100.)

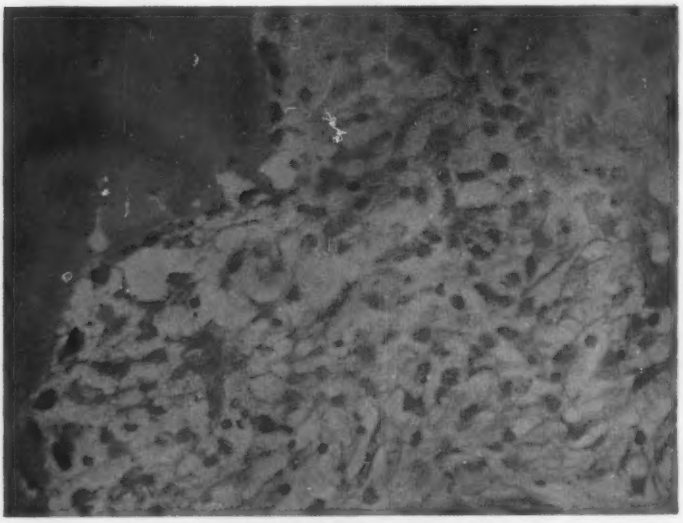

FIG. 17

Photomicrograph from the same area, to show cortical bone lined with osteoblasts and " foam cells." (Haematoxylin and eosin, ×400.)

surrounding cleft-like spaces. The appearances suggest phagocytosis of lipoids, which have produced a fairly marked outgrowth of the fibrous stroma.

" This suggests continued activity of the lipoidosis of Hand-Schüller-Christian disease."

Haematological studies—The blood picture remained normal except for a moderate hypochromic anaemia and a raised erythrocyte sedimentation rate varying between 40 millimetres/first hour and 65 millimetres/first hour—a typical "blood picture" being: polymorphonuclear leucocytes 80 per cent; lymphocytes 13 per cent; monocytes 5 per cent; eosinophils 1 per cent; reticulocytes 1 per cent.

Sternal marrow biopsy (November 1951)—"The marrow is reactive—normoblastic and leucopoietic. There are considerable numbers of typical plasma cells diffusely scattered throughout the haemopoietic islets, but no appearance which would indicate the presence of plasmocytoma in this biopsy. There is no indication of lipoidosis. There is an eosinophil myleocytic predominance compatible with a peripheral eosinophilia."

Blood chemistry—Serum calcium, inorganic phosphates, phosphorus and cholesterol were always within normal limits.

Endocrine system—There was no evidence to suggest any endocrine dysfunction, such as diabetes insipidus or hypogonadism.

TREATMENT 1951-1955

Immobilisation in a plaster-of-Paris posterior shell was carried out with fixation of the patient's right humerus in a plaster arm extension appliance. A high protein diet with supplementary vitamins was given with oral iron therapy.

Radiotherapy—Because of the lack of much response to the original course of x-ray therapy in 1949, Professor R. McWhirter, considered that further treatment of this nature was not to be recommended. However, in 1953 a course of nitrogen mustard was tried over a period of two months, but no obvious change in condition was noted.

Cortisone therapy—After the episode of multiple pathological fractures in 1951 and because of her poor general condition with cachexia, low grade pyrexia and a raised sedimentation rate, cortisone was given orally for three weeks. The rationale for this form of therapy was the eosinophilic character on biopsy and the observation by Donohue *et al.* (1950) of some temporary improvement in cases of eosinophilic leukaemia when treated with A.C.T.H. therapy. However, although the general condition of the patient improved, there was no cellular alteration in the pathology on biopsy and any improvement was most probably due to the immobilisation.

Operative treatment—In November 1954 it was decided that because there had been no evidence of bone repair in any of the lesions except in the fractured humerus, any attempt to alter the balance in favour of bone regeneration was justified. Hence radical saucerisation of the diseased area in the region of the pathological fractures in both femora was carried out and the areas were packed with cancellous bone from the bone bank. After six months' immobilisation, although there was little evidence of new bone formation, an attempt to mobilise the patient was decided upon. Following physiotherapy, bilateral non-weight-bearing walking calipers were tried, and walking with crutches finally achieved.

DISCUSSION

Etiology and pathogenesis—This condition has been defined by Jaffe and Lichtenstein as a "specific inflammatory histiocytosis, whatever its etiological agent may ultimately prove to be." It is regarded as being primarily a disease involving the reticulo-endothelial system and this accounts for its diffuse nature in both skeletal and extra-skeletal tissues, but it is still classified under the heading of "diseases of uncertain nature" as a non-lipoid histiocytosis by Payling Wright (1953). This also includes such a condition as sarcoidosis. It has been suggested as resulting from an allergic response to an unknown agent and the early clinical features of fever, raised sedimentation rate and toxaemia with the local and peripheral

eosinophilia, areas of focal necrosis, etc., may support this concept. The appearance of "foam cells" as seen in this case in the early stage could represent the usual phagocytosis by the histiocytes of degenerative debris, resulting from the necrotic process of the primary condition. It is suggested that the unknown agent could be an abnormal protein which has been produced by the reticulo-endothelial cells in place of the modified plasma proteins or antibodies which are normally produced by this system. It is not truly a generalised condition in the skeletal system as it is limited to the membrane bones and the proximal aspect of the limb bones only, and this is clearly seen in this case (Fig. 18). This uncertainty of localisation could be explained by our lack of knowledge of the anatomy of the reticulo-endothelial system in bone.

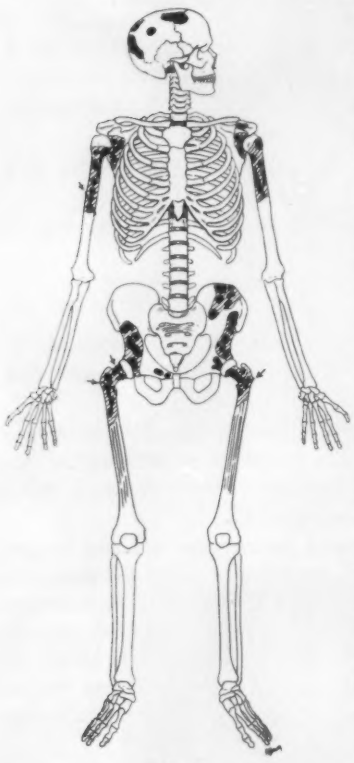

FIG. 18

Skeletal diagram to show diseased areas (in shading) with cyst-like translucent areas (in black), in the membrane bones and proximal part of the limb bones.

In the later stage of this condition, which is termed Hand-Schülier-Christian disease or lipoid granulomatosis or chronic disseminated histiocytosis (Lichtenstein 1953), the predominant cell type is now a "foam cell" or a lipoid histiocyte with the granulomatous background. The "allergic" picture of eosinophilia, focal necrosis, etc., has now disappeared. This suggests that the disease process has changed and that it is now some metabolic disturbance of the usual synthesis and storage of certain lipoids—in particular the phospho-lipoid lecithin. Because of this, there has been a tendency to group this later stage of histiocytic granulomatosis with other true lipoid storage diseases, such as xanthoma, Gaucher disease in which the lipoid is a cerebroside, Nieman-Pick disease, Tay-Sach disease, etc. Jaffe (1938) considers that the naming or differentiation of these varied conditions should be based upon the organs affected, the chemical nature of the lipid and the presence or absence of excessive amounts of the lipid in the blood. But with these latter conditions, the metabolic disorder is the primary manifestation and is not, so far as is known, the later stage of any primary disease process, therefore the condition of histiocytosis should not be included in this group.

The distribution in this case is of interest in that there is involvement of both skeletal and extra-skeletal tissues. The latter is represented by the typical changes in the lung pattern due to a histiocytic proliferation of the normal septum cells found in the pulmonary interstitial structures, of the blood-borne macrophages and the reticulo-endothelial cells in the local connective tissue. The anatomy of the reticulo-endothelial system in the respiratory system has been recently reviewed by Payling Wright (1953). Lichtenstein emphasises how clinical episodes resulting from spontaneous pneumothorax, with honeycombing of the lungs, and pulmonary fibrosis, are the "rather common hallmark" of chronic disseminated histiocytosis of Schüller-Christian disease even in the absence of obvious skeletal defects. This radiological appearance in this case simulates the condition of "eosinophilic lung" which is an acute allergic respiratory condition seen commonly in the Far Eastern countries. During the last six years there has been no obvious change in the lung field pattern and never any episodes of spontaneous pneumothorax nor other respiratory complaints noted. There are no other areas of soft tissue involvement in the spleen, liver, lymph glands, skin or more importantly

in the cranio-hypophysial region to produce the endocrine dysfunction of diabetes insipidus or hypogonadism, etc., which one would expect with such marked calvarial involvement. The exophthalmos recorded in this case is probably coincidental, as it was noted by the patient several years before the onset of any symptoms referable to the present condition.

The normal blood chemistry is in keeping with the described condition and is one of the factors in differentiating this lesion from a truly lipoid storage disease.

Change in cellular pattern—In this case there is presented pathological evidence from biopsies carried out chronologically of the transformation from a histiocytic granulomatous stage with predominantly eosinophils to the lipoid granulomatous stage with predominantly " foam cells," during the past six years. This is in support of Farber, and Green and Farber's concept of a conversion or change of an eosinophilic granuloma to a lipoid granulomatosis. It also indicates how the pathology at a given time refers to one particular stage of one condition (Engelbreth-Holm), or represents " closely related expressions of a single nosologic entity " (Lichtenstein).

The cellular change associated with repair or healing such a fibrosis, bone regeneration, etc., although seen in the pathological fracture of the humerus, was not seen on radiology or from the biopsies in any other diseased area. In 1954 the last biopsy showed some evidence of osteoblastic activity and the production of a fibrous stroma, but this specimen was taken from a fracture site and therefore is not necessarily indicative of a generalised tendency towards healing. Indeed the healing of the fractured humerus, which was oblique in nature and therefore presented the greatest surface and stimulus to bone regeneration, must be regarded as an exceptional change in the pattern of the disease process in this patient.

It is difficult to envisage bone regeneration through these gross areas of lipoid granulomata with its poor vascularity without the stimulus of some form of trauma. The numerous reports of complete disappearance of involved areas and the healing of pathological fractures (Versiani *et al.* 1944, Hill 1949, and Ponseti 1948) originate mainly from the young patient in whom the stimulus to repair and growth is marked.

Treatment—Radiotherapy was given to this patient in the early stage with temporary alleviation of pain which recurred within six months. There are many varied reports of the favourable effects of this form of treatment by Ponseti (1948), and by Childs and Kennedy (1951), etc., as well as its being of little value, or having little effect (Hamilton *et al.* 1946). Lichtenstein rightly points out the difficulty of evaluating such treatment for such a condition with its varied expressions of stage, severity, distribution and age of patient. He also suggests that roentgen therapy may accelerate the change from the eosinophilic stage to that of a "lipoid granuloma." This could not be evaluated in this case, except that from biopsy before, the picture was of a histiocytic granuloma with numerous eosinophils, and eighteen months later it was one of a lipoid granuloma.

The cortisone therapy did not appear to have improved the patient clinically or to have changed the cellular pattern, but it should be noted that it was not given during the acute phase, in 1949, when the eosinophilic picture predominated. Independently of this case, Blahd *et al.* (1951) recorded the improvement of a case of Hand-Schüller-Christian disease which had responded to cortisone.

In this case the more rational approach was in deciding to operate and to excise radically the diseased area and pack with cancellous bone chips from a bone bank. Although there has been no obvious radiological evidence of bone regeneration, movement at both hip joints is now possible without pain or muscle spasm. A pseudarthrosis appears to have been produced. This patient is now walking with crutches and bilateral walking calipers. It is suggested that in adult cases of this condition, in any area which has failed to show evidence of bone regeneration or repair, especially in a fracture site of a limb bone, radical saucerisation and bone grafting should be considered.

Prognosis—This must, of necessity, be guarded in this case as well as for the condition generally, especially with the multiple involvement of skeletal and extra-skeletal systems. With the involvement of the lungs, intercurrent infection, pneumothorax, cardiac failure could occur and kill. It is commonly stated that without complications this condition does not end fatally. But because of the variation of severity, the age group involved and the pathology, a prognosis cannot be accurately stated, and indeed this is in keeping with the condition generally in which even a diagnosis of certainty cannot be given until the case has been observed over years with repeated pathological studies.

We are indebted to Dr R. F. Ogilvie, Pathologist to the Royal Infirmary of Edinburgh, for his consideration and report on the pathological material in this case.

Part of this work was carried out while one of us (R. B. D.) was holding a Research Fellowship of the Scottish Hospitals Endowment Research Trust.

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ANEURYSMAL BONE CYST

A Report of Three Cases

F. W. TAYLOR, SHEFFIELD, ENGLAND

Aneurysmal bone cyst was first described as a distinct entity by Jaffe and Lichtenstein in 1942. Further cases have since been reported in America (Mayer and Kestler 1944, Jaffe 1950, Lichtenstein 1950, 1953, Besse *et al.* 1953), but so far as can be ascertained none has been reported in this country.

The three cases described here are considered to be examples of this condition.

CASE REPORTS

Case 1—A girl aged sixteen was medically examined for acceptance as a student nurse. She had been in good health but stated that for about six months she had noticed a dull pain in the lower back, especially after standing for more than a short time. On examination there

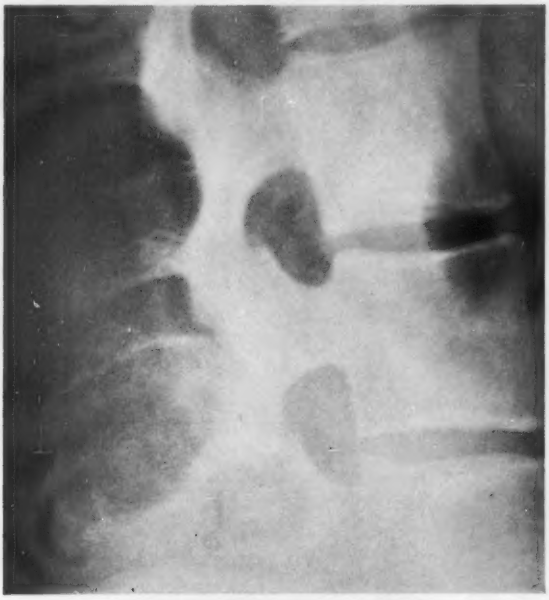

FIG. 1
Case 1—Cystic expansion of the spinous process of the third lumbar vertebra.

was a slight prominence of the spinous process of the third lumbar vertebra, which appeared to be broader than its fellows. There was slight limitation of flexion of the lumbar spine, but other movements were full.

Radiographic examination showed a lesion involving the third lumbar spinous process, which had lost its normal contours and was expanded to about two or three times its normal size. The process appeared to be limited peripherally by only a thin shell of bone (Fig. 1). Radiographs of the rest of the skeleton showed no other lesion. There was no abnormality in

T2(8)

the blood chemical examinations. While the girl was under observation the discomfort in the back became more severe and the expansion of the spinous process of the third lumbar vertebra became more obvious. It was decided to explore and if possible remove the mass.

Operation—A mass about 8 centimetres long and 5 centimetres broad involved the spinous process of the third lumbar vertebra and extended over the spinous processes of the second and fourth lumbar vertebrae. The mass was removed completely after dividing both laminae of the second, third and fourth lumbar vertebrae.

Progress—When last seen one year after the onset the patient was free from symptoms and had a full range of movements of the spine. Radiographs showed no evidence of recurrence.

Pathological examination—The specimen consisted of a bony mass measuring $8 \times 5 \times 5$ centimetres (Fig. 2). The outer surface was smooth and well defined. Cross-section revealed a surface dark red in colour, which was honeycombed with numerous cysts, varying from

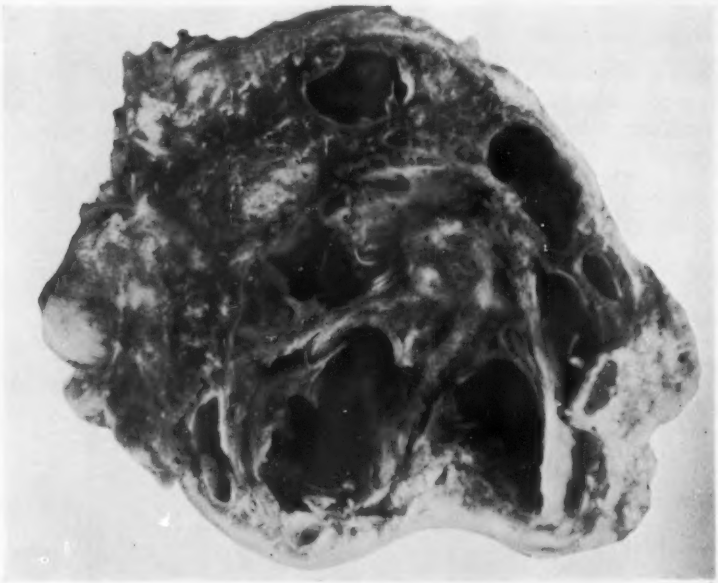

FIG. 2

Case 1—Cross-section of the specimen, showing the sponge-like appearance with cystic spaces.

pinhead size to a centimetre or two in diameter. Most of the cysts contained blood or organising thrombi. The cystic spaces were separated by septa of varying thickness, some containing thin shell-like plates of bone and others consisting only of fibrous tissue.

On microscopic examination, the cysts were seen to be lined in places by flattened cells, but these were not always apparent. Cancellous bone was abundant in the septa, which also contained varying amounts of fibrous tissue and numbers of multinucleated giant cells (Figs. 3 and 4).

Case 2—A girl aged thirteen complained of aching in the left elbow for seven weeks. For the previous two weeks the elbow had been swollen. On examination there was a warm swelling around the elbow with slight restriction of movement. Radiographs showed a small cystic lesion in the lower end of the humerus (Fig. 5). It was considered that this was possibly a tuberculous infection of the elbow and the arm was immobilised in plaster. Further radiographs showed a rapid increase in the size of the lesion (Fig. 6).

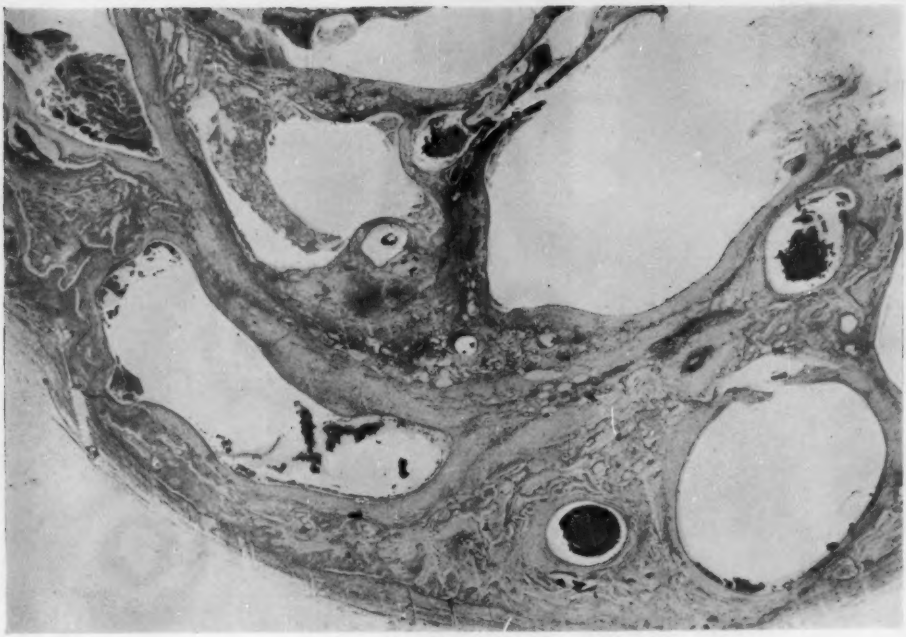

FIG. 3

Case 1—Several cystic spaces are shown. Note the smooth lining and thrombi in some spaces. (H. and E., ×7.)

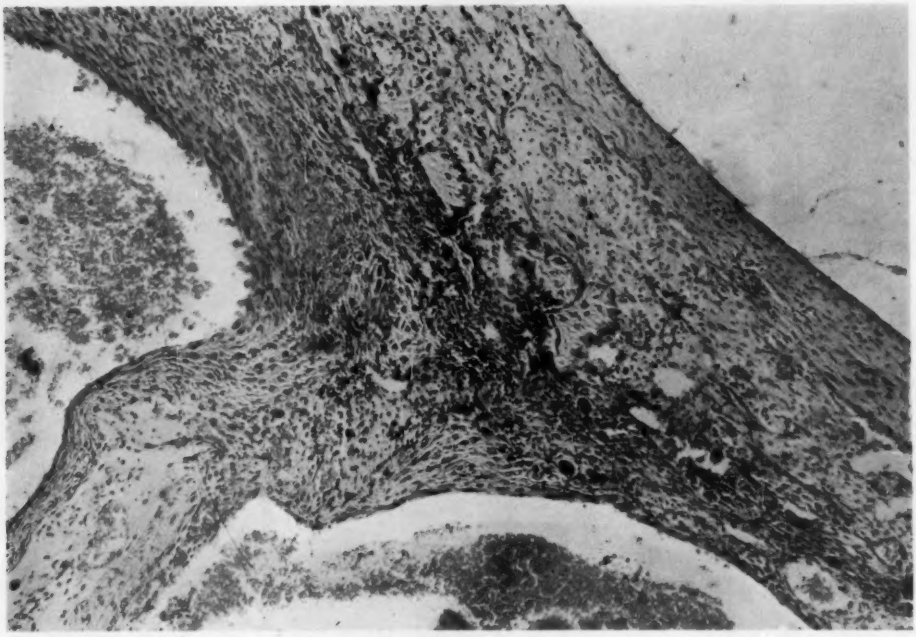

FIG. 4

Case 1—Showing the cancellous bone; flattened cells lining the cystic spaces; stromal cells and multinucleated giant cells. (H. and E., ×7.)

FIG. 5

FIG. 6

Case 2. Figure 5—Early radiograph showing lesion in lower end of humerus. Figure 6—Four months later, showing the increase in the size of the lesion. Note the trabeculation and eccentric expansion, well shown in the lateral view.

FIG. 7

FIG. 8

FIG. 9

Case 2. Figure 7—Radiograph eight months after first examination and six weeks after biopsy, showing apparent consolidation of the lesion. Figure 8—Eleven months later, four months after x-ray therapy, showing loss of the cystic appearance and recalcification of the lower humerus. Figure 9—Two years later still.

Biopsy—At operation the lower end of the humerus was exposed. The surface of the bone had a bluish-black appearance and the cortex was easily cut with a scalpel. The cyst contained nothing but blood. Sections of the small piece of cortex showed flattened cells lining the inner surface, areas of osteoid tissue and small cystic areas containing blood.

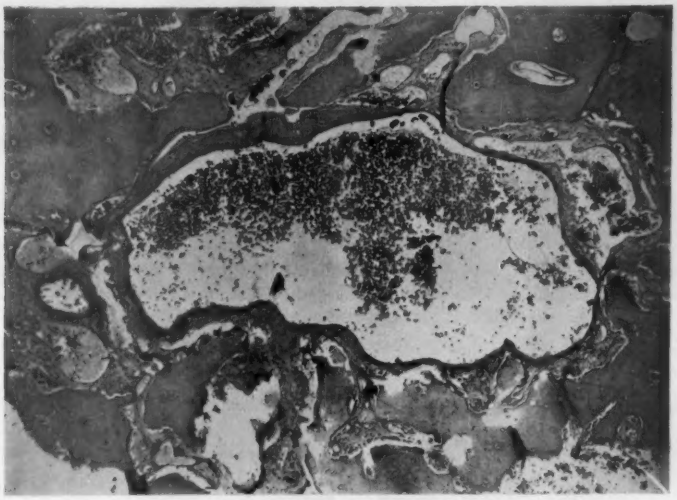

FIG. 10
Case 2—Showing bone, cystic spaces and flattened lining cells. (H. and E., ×35.)

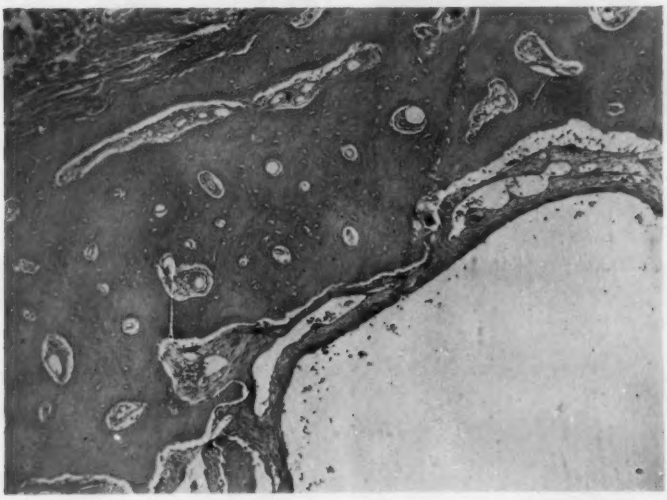

FIG. 11
Case 2—Cancellous bone; the margin of a large cyst with flattened cells lining it.

It was thought at the time that the lesion was probably an osteoclastoma, but a review of the sections suggests that the appearances are consistent with a diagnosis of aneurysmal bone cyst (Figs. 10 and 11).

Progress—Although no attempt had been made to remove the bulk of the lesion at biopsy,

FIG. 12
Case 3—Initial radiograph showing cystic expansion of spinous process of the atlas.

FIG. 13

FIG. 14
Case 3. Figure 13—Three months later. Increase in size of the lesion. Figure 14—Lesion apparently increasing in size after x-ray therapy.

FIG. 15

FIG. 16
Case 3. Figure 15—Five months later. Recalcification of the margin of the cyst. Figure 16—Eight years after onset. Spinous process still larger than normal but well consolidated.

it appeared to settle down and radiographs showed calcification of the cystic area (Fig. 7). A course of deep x-ray therapy was given and the patient has since been completely free from symptoms. When she was last seen three years after the first examination there was a full range of movements at the elbow and radiographs showed no sign of recurrence (Figs. 8 and 9).

Case 3—A girl aged four had had pain in the neck for about three months, with some limitation of movement of the head. On examination a palpable swelling was noted immediately below the occiput. Movements of the head, especially dorsiflexion, were limited. Radiographs showed a cystic expansion of the spinous process of the atlas. The outline of the expanded process appeared to consist of a thin shell of bone (Fig. 12). The discomfort increased and further radiographs showed a considerable increase in the cystic expansion of the spinous process (Fig. 13).

Treatment and progress—It was decided that operation was inadvisable and a course of deep x-ray therapy was given. The discomfort gradually disappeared and the child regained a full range of neck movements. She has remained free from symptoms over the past nine years. Radiographs taken two weeks after the x-ray therapy showed an apparent increase in the size of the cyst (Fig. 14) but this was followed by a gradual loss of the cystic appearance, and the spinous process, though larger, has remained well calcified (Figs. 15 and 16).

DISCUSSION

Similar lesions have been noted in most parts of the skeleton but they have usually been thought to be variants of tumours such as osteoclastoma (Ewing 1930). Geschickter and Copeland (1949) described a similar lesion as a borderline between osteoclastoma and osteitis fibrosa. Most of the reported cases have been in young people—usually under the age of twenty. Pain, which is not usually severe, is the commonest symptom (Besse *et al.* 1953). There may be limitation of movement when the lesion is close to a joint, as in Case 2. The lesion can occur anywhere in the skeleton but it appears to favour the long bones and the vertebrae (Lichtenstein 1950, Besse *et al.* 1953). In the published cases there is no record of local invasion or of metastases, but the lesion may cause damage by local pressure and in spinal lesions there is a danger of cord compression (Besse *et al.* 1953). The prognosis is good, and it has been stated (Lichtenstein 1953) that curettage, even if incomplete, may cure the condition. X-ray therapy is said to be effective and it is recommended for tumours in inaccessible positions (Lichtenstein 1953). The third patient reported here has remained free from symptoms for nine years after deep x-ray therapy.

The radiographic features of an eccentrically placed expanded cyst-like lesion with its margin outlined by a fine rim of bone are well shown in the cases reported here (Figs. 1, 6, 12). The main pathological characteristics as described by Lichtenstein (1950) are also well illustrated in the specimen removed from Case 1 (Fig. 2). The normal architecture of the bone is completely transformed, the cross-section showing a spongy appearance with blood-filled spaces of varying size. The larger spaces are separated by septa which often contain thin plates of bone, but the smaller spaces are commonly separated only by thin fibrous septa. Microscopically, the cystic spaces are bordered by supporting connective tissue which often shows evidence of organising haemorrhage, multinucleated giant cells and new bone formation (Figs. 3, 4, 10, 11). Despite their large size the vascular channels do not appear to have fibromuscular or elastic walls (Lichtenstein 1950).

Jaffe, who recorded twenty cases in 1950, was reluctant to accept the lesion as a separate entity, and regarded it as a modification of some pre-existing tumour. It is difficult to concur with this view because in other published cases, as in Cases 1 and 2, there was no evidence of any other recognisable tumour. Ewing's (1928) opinion that it represented an osteoclastoma, modified by free communication with large blood vessels, is also difficult to accept. It is unlikely that a tumour that exhibits none of the histological features of an osteoclastoma (apart from the presence of small numbers of multinucleated giant cells), which occurs in a

different age group (under twenty rather than over twenty), and which has never been shown to metastasise, can belong to the same group. Furthermore several authors have noted that even incomplete removal will often cure the condition (Lichtenstein 1950, 1953, Besse *et al.* 1953). Such behaviour is not characteristic of osteoclastoma.

Confusion with simple bone cysts, or with osteitis fibrosa cystica, should not occur because such cysts do not exhibit the same vascularity or indeed contain blood, except in the presence of a fracture.

Although I agree with Lichtenstein that this is most probably a distinct pathological entity, I believe that the term aneurysmal bone cyst is inappropriate. An aneurysm has been defined as a space or sac formed by the widening or extension of the lumen of an artery, and thus containing blood or clot. The lesion in question certainly does not fit into this definition.

In studying Cases 1 and 2 and other published cases, I have seen no reason to suggest that these vascular lesions with endothelial-lined spaces should not be considered to be a type of cavernous haemangioma. Similar views were expressed by Hadders and Oterdoom (1956) at a recent meeting of the Pathological Society at the Royal College of Surgeons. It is suggested that the name " haemangiomatous bone cyst " is preferable to " aneurysmal bone cyst."

SUMMARY

1. Three cases of aneurysmal bone cyst are recorded. It is suggested that the condition is a type of cavernous haemangioma and the term haemangiomatous bone cyst is preferred.
2. A survey of the published cases suggests that in its early stages the lesion is amenable to local surgery, which is the treatment of choice, but for inaccessible lesions deep x-ray therapy may be effective.

I am indebted to Mr A. Dornan for permission to publish Case 1, to Mr F. W. Holdsworth for Cases 2 and 3, and to Dr J. L. Edwards for advice on the pathological material.

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ANEURYSMAL BONE CYST

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Aneurysmal bone cyst is a benign lesion of bone. It was first described by Jaffe and Lichtenstein in 1942 and consists essentially of fibrous tissue honeycombed by vascular spaces. The lesion has been observed in many parts of the skeleton but it occurs especially in the shafts of long bones and in the vertebral column. It causes localised distension and destruction of the affected bone, limited peripherally by a thin bony shell (Fig. 1).

In the past the lesion has usually been regarded as an atypical giant-cell tumour or a cavernous haemangioma of bone. The purpose of this paper is to support the contention of Lichtenstein (1950) that aneurysmal bone cyst is a clinical and pathological entity, which deserves wider recognition than it has had hitherto.

The case reports that follow illustrate some of the more important features of the lesion.

CASE REPORTS

Case 1 (Reported by the courtesy of Mr D. Lloyd Griffiths)—A girl of thirteen complained of a painful swelling of the upper end of the right ulna of three months duration, which had increased rapidly in size. There was no history of injury. Examination revealed a hard swelling arising from the upper end of the ulna, which had a nodular surface. The swelling was warmer than the surrounding tissues, but there was no pulsation. Radiography disclosed a cyst arising from the anterior surface of the upper third of the ulna. It extended from the base of the coronoid process for 6½ centimetres down the shaft of the bone (Fig. 1). The cyst had destroyed the anterior cortex of the ulna and invaded the medullary canal; it was bounded anteriorly by a thin bony shell.

Operation (22.3.49)—The cyst was excised together with the upper end of the ulna.

Histology—Pathological examination revealed numerous large cystic spaces containing fresh blood (Fig. 2). The walls of the cyst were lined by cellular connective tissue in which there were numerous giant cells.

Progress—The head of the radius dislocated anteriorly, and an attempt to retain the reduced position was unsuccessful. Later the upper end of the ulna was replaced by an acrylic prosthesis. The patient has now good function in the arm and is training as a nurse. There has been no recurrence of the cyst.

Case 2—A girl of seventeen felt a sharp pain in the lower part of the back while pushing a door against resistance. The pain persisted until her admission to hospital two years later; it sometimes radiated down the posterior aspect of the thigh to the leg, and was aggravated by coughing, sneezing and exercise. Clinical examination revealed restriction of flexion and right lateral bending in the lumbar spine and tenderness to the right of the lumbo-sacral joint. Radiographs disclosed destruction of the spinous process, lamina and right superior and inferior articular processes of the fifth lumbar vertebra. There was also erosion of the transverse process and the adjacent part of the body of this vertebra, and of the upper part of the ala of the sacrum on the right side (Figs. 3 and 4).

Biopsy revealed what was thought to be a vascular " tumour " lying deep to erector spinae, and later reported to have the histological features of an aneurysmal bone cyst.

Operation (19.3.54)—The "cyst" was exposed; it was deep reddish brown in colour, and well demarcated from the surrounding tissues. Almost the whole of the neural arch of the fifth lumbar vertebra was destroyed, and only the tip of the spinous process remained, loosely attached to its surface (Fig. 5). The cyst contained friable vascular tissue which was removed partly by curettage and partly by suction. The dura was now exposed and the fifth lumbar nerve root could be seen emerging from it on either side. The bone in the floor of the cyst was smooth; its appearance suggested erosion by pressure rather than destruction by tumour (Fig. 6).

Histology—The specimen consisted of many fragments of soft, highly vascular, reddish stranded connective tissue. Microscopy showed fibrous tissue, in some areas dense and collagenous, but for the most part cellular and with strands of osseous metaplasia. Running throughout were wide vascular channels containing fresh blood, except for a few where newly formed thrombi were found. Giant

FIG. 1

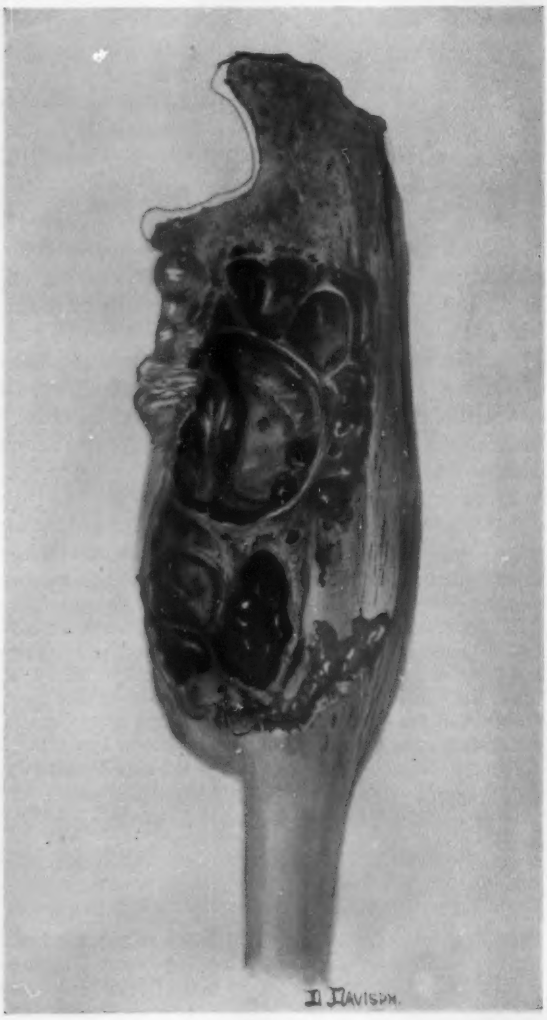

FIG. 2

Case 1. Figure 1—Radiograph of a typical lesion in the upper end of the ulna (Case 1). The cyst has caused marked " ballooning " of the bone, and is bounded anteriorly by a thin shell of subperiosteal bone. Figure 2—Section through the lesion showing gross appearance.

cells were numerous, especially at the borders of the trabeculae, and tended to be smaller in size than those in a giant-cell tumour. The appearances were those of aneurysmal bone cyst.

Progress—The patient was examined fifteen months after operation. She then complained of slight aching pain in the lower part of the back after exercise. There was free movement in the lumbar spine. The radiograph showed normal density of the ala of the sacrum, and partial regeneration of the neural arch. The right transverse process and body of the fifth lumbar vertebra had reossified (Fig. 7).

Case 3 (Reported by the courtesy of Mr J. E. Paterson)—A man of twenty complained of left-sided sciatica. The pain commenced after an injury at rugby nine months earlier, and for the past month had been severe enough to compel rest in bed. Clinical examination revealed guarded movement of the lumbar spine and tenderness over the left sacro-iliac joint. Lasègue's sign was positive at

FIG. 3

Case 2—A lesion of the fifth lumbar vertebra and sacrum as seen from behind. Almost the whole of the neural arch, and part of the body and right transverse process of the fifth lumbar vertebra, are destroyed.

FIG. 4

Case 2—An oblique radiograph to demonstrate the destruction of the neural arch of the fifth lumbar vertebra and the cyst in the right ala of the sacrum.

FIG. 5

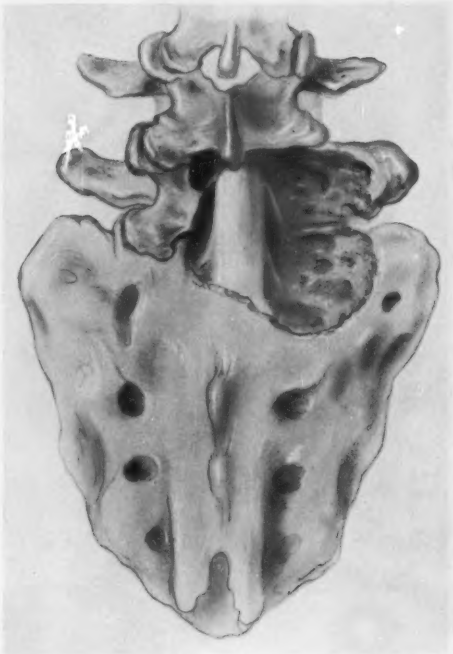

FIG. 6

Case 2—Diagrams showing the appearance of the cyst at operation (Fig. 5) and the degree of bone destruction (Fig. 6). The dura is exposed in the floor of the cyst.

FIG. 7

Case 2—A radiograph three months after operation, for comparison with Figure 2. There is evidence of healing. The lesion in the transverse process of the fifth lumbar vertebra is now more obvious due to the calcification of its bony shell.

5 degrees on the left side and at 45 degrees on the right. The left ankle jerk was depressed. There was no other abnormal neurological sign. No lesion was observed in the radiographs, and the pre-operative diagnosis was a lumbo-sacral disc protrusion.

Operation (27.10.54)—When the muscles were being stripped from the spinous processes a large cyst arising from the upper part of the sacrum was opened. Its posterior wall was no thicker than an egg shell, and it was lined by a thin layer of vascular tissue which peeled easily from the sides of the cavity. The contents of the cyst were not removed completely, but its deeper part probably involved the dura.

Progress—The patient complained of severe pain in the leg for the first few days after the operation, but thereafter his convalescence was uneventful. When discharged from hospital three weeks later he was free from pain and walking in comfort. A further study of the pre-operative radiographs disclosed destruction of the upper part of the sacrum on the left side.

FIG. 8

Case 3—Cyst in the left ala of the sacrum eight months after operation. Its outline is much clearer than in the pre-operative radiograph, due to the calcification of the bony shell.

The patient was re-examined eight months after the operation. He then complained of a little weakness of the back when lifting heavy weights. There was a good range of movement in the spine, and Lasègue's sign was negative. A radiograph showed commencing reossification of the cyst in the left ala of the sacrum (Fig. 8).

Case 4—A boy of eight received a kick on the left leg three weeks before admission to hospital. A swelling appeared, but was not painful. Examination revealed a firm non-tender fusiform swelling of the lower third of the tibia. A radiograph disclosed a cystic lesion of the lower part of the shaft of the tibia, which had produced some expansion of the bone (Fig. 9). The cyst was finely trabeculated and had a thin bony shell.

Operation and progress—The cyst was opened and contained bloodstained fluid, and a thin lining membrane which was not removed completely. The patient was traced fourteen years later; he had no complaints. A radiograph showed a small cyst in the lower part of the tibia but the rest of the bone was normal in appearance. The sections were re-examined recently and are typical of an aneurysmal bone cyst.

FIG. 9
Case 4—Radiograph of a lesion of the left tibia. The uniform expansion of bone and the degree of involvement of the medullary canal are unusual.

Case 5—(Reported by the courtesy of Mr E. W. Somerville)—A boy of fourteen complained of pain in the right leg of six months duration, and of a swelling which had appeared some time later. There was no history of injury. Examination revealed a tender swelling of the postero-medial aspect of the upper third of the tibia. Radiographs disclosed a cyst which had caused a localised expansion of the posterior cortex of the tibia, bounded posteriorly by a thin shell of subperiosteal bone (Fig. 10). The cyst extended within the medulla to the upper tibial epiphysis.
Biopsy—The cyst was lined by vascular osseous tissue. Histological examination showed connective tissue with numerous haemorrhages, multinuclear giant cells, and fragments of bone. The fluid from the centre of the cyst contained red blood cells and a few leucocytes. The diagnosis was a haemorrhagic bone cyst of tibia.
Treatment and progress—The cyst was re-explored on 21.8.55 and the lining carefully removed by curettage. The cavity was packed as far as possible with slivers of bone taken from the opposite tibia. The post-operative progress was satisfactory. Radiographs showed progressive consolidation of the grafts, and no extension of the cyst.

Apart from the contributions of Jaffe (1950) and Lichtenstein (1950, 1953) there are as yet only a few isolated case reports of aneurysmal bone cyst in the surgical literature. This may be explained partly by lack of familiarity with the clinical and radiographic features of the disease, and partly by the rarity of the lesion. The account which follows is based on a

review of the relevant literature, and reports of atypical giant-cell tumours which may now be reclassified as aneurysmal bone cysts.

CLINICAL FEATURES

Age and sex incidence—Most of the patients are children, adolescents, or young adults. Both sexes are affected, males more frequently than females.

Distribution—There is a preference for the vertebral column and the shafts of long bones. The lesion occurs at all levels in the spine. The neural arch is involved oftener than the body of a vertebra. In the long bones the cyst is usually situated beneath the periosteum of the shaft and rarely invades the epiphysis. Aneurysmal bone cysts have also been described in most of the flat bones, skull, and bones of the hands and feet. Contiguous bones are often affected, especially in lesions of the spine (Figs. 3 and 4).

Symptoms—Pain as a rule is not severe, but it may be if the collapse of a vertebra has caused pressure on a nerve root (Case 3). The symptoms often date from a specific injury. This may have drawn attention to a pre-existing lesion, but this is not always so, for sometimes a radiograph taken shortly after injury was negative.

Clinical signs—When the cyst is in the spine there is always some restriction of movement. It may be considerable if there is destruction and collapse of a vertebra, when paraplegia is a possible complication. In the long bones the swelling is often palpable; it may become very large and interfere with the function of the limb.

FIG. 10

Case 5—Radiograph of a typical lesion of the right tibia. Note the "ballooning" of the posterior cortex.

RADIOGRAPHIC FEATURES

In a long bone the lesion appears as a cyst arising from the cortex of the shaft, and limited peripherally by a thin shell of subperiosteal bone (Fig. 1). There is usually some destruction of the underlying cortex, but the cyst does not as a rule invade the medulla, and only rarely is there uniform expansion of the affected bone (Fig. 9). The cyst frequently has a mottled or trabeculated appearance, and there may be irregular strands of ossification which are sometimes a feature of older lesions. In the spine the picture is often obscure, because it may be difficult to demonstrate the bony shell, and the osteolytic process is then likely to be attributed to a primary or metastatic tumour (Figs. 3 and 4). These tumours are, however, usually situated in the body of a vertebra, whereas an aneurysmal bone cyst is found more frequently in the neural arch or transverse process.

PATHOLOGY

When the bony shell of the cyst is opened one is confronted by strands of deep reddish brown vascular tissue honeycombed by spaces filled with blood (Fig. 2). Blood wells out of the cavity, indicating a free circulation within the cyst, but there is seldom any pulsation

or brisk arterial bleeding. Small strands of fibre bone may be interspaced throughout the tissue and give it a gritty feel. Sometimes the tissue is less vascular and has a greyish appearance; it is then distinctly more collagenous, suggesting an attempt at repair.

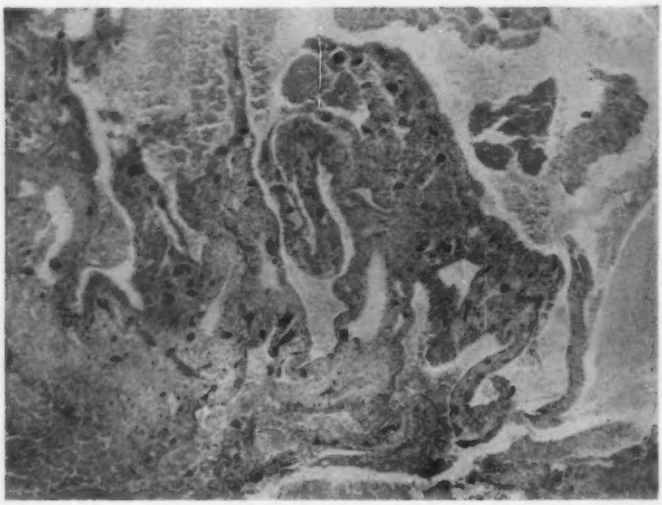

FIG. 11
Section showing the topography of the lesion. There are many large vascular spaces separated by strands of fibrovascular tissue. (H. and E., ×45.)

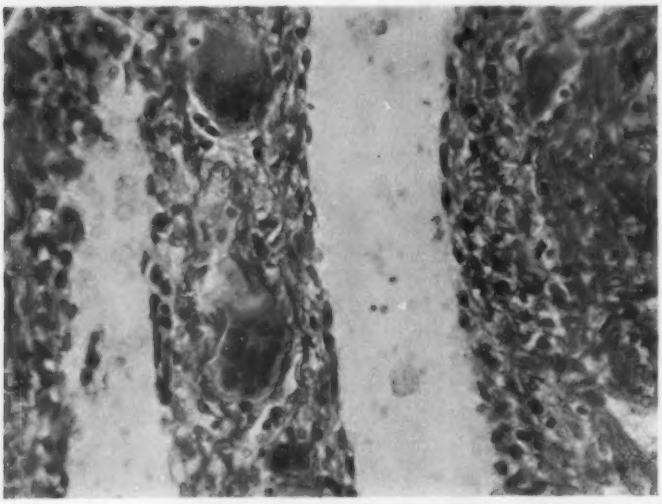

FIG. 12
Section showing strands of fibrovascular tissue containing multinucleated giant cells. (Masson stain, ×270.)

In a typical lesion histological examination reveals large vascular spaces separated by strands of fibrous tissue in which there are numerous capillaries (Figs. 11 and 12). Multi-

nucleated giant cells are common around areas of haemorrhage; these giant cells are smaller and have fewer nuclei than those of a giant-cell tumour. Filaments of osteoid tissue and new bone are often present in the fibrous tissue (Fig. 13).

FIG. 13

Two large vascular spaces are seen in the upper part of the field. The tissue lining the space on the left contains numerous giant cells. In the lower part of the field there are filaments of osteoid tissue surrounded by osteoblasts. (H. and E., ×58.)

DIAGNOSIS

The lesion which has been described has long been recognised. Ewing (1940) referred to it as an aneurysmal giant-cell tumour, and Coley (1949), Thompson (1954) and others as an atypical or subperiosteal giant-cell tumour. Apart from the presence of giant cells, aneurysmal bone cyst has little resemblance to a typical giant-cell tumour. The main points of difference between them are indicated in Table I.

TABLE I

COMPARISON OF ANEURYSMAL BONE CYST AND GIANT-CELL TUMOUR

	Aneurysmal bone cyst	Giant-cell tumour
Age	Usually children and young adults	Patients usually over thirty years
Site	Shafts of long bones and vertebrae (neural arch). May involve contiguous bones	Epiphysial end of long bones especially upper end of tibia and lower end of femur
Appearance	In long bones is usually situated eccentrically beneath periosteum	Expands bone from within
Pathology	Fibrovascular tissue honeycombed by vascular spaces. Giant cells small and not present throughout tissue	Large numbers of multinucleated giant cells in vascular spindle-celled stroma
Behaviour	Strong tendency to spontaneous regression. Incomplete removal often followed by cure	Invasive and aggressive. Sometimes malignant from onset

A simple or unicameral bone cyst is situated in the centre of the metaphysis, and seldom causes pain unless there is a spontaneous fracture. It contains yellowish fibrous tissue which

is firmly attached to the walls of the cyst, and amber coloured fluid which is not bloodstained unless there has been a recent fracture.

A number of aneurysmal bone cysts have also been incorrectly diagnosed as haemangiomas. A true angioma is situated within the medulla of a long bone, or the vertebral body. It produces a cystic lesion of the bone with a marked " soap bubble " effect. Microscopic examination reveals a large number of dilated capillaries lined by endothelium in a loose connective tissue stroma; the formation of osteoid tissue and new bone is not a feature.

When the lesion is in the shaft of a long bone it may have some resemblance to an osteolytic osteosarcoma or a malignant bone aneurysm. Both these tumours are intensely painful; the area of bone destruction is not so well defined, nor is there the trabeculation of an aneurysmal bone cyst, and the tumour quickly invades the soft tissues and destroys the bony shell.

TREATMENT

All case reports agree that the lesion is benign. Lewis (1924) reviewed seventeen giant-cell tumours of the spine, of which many might now be reclassified as aneurysmal bone cysts. Recovery occurred spontaneously, or after partial removal of the tumour, in thirteen of the seventeen patients. Lewis stated that he had doubt as to the neoplastic nature of some of the tumours because of their clinical behaviour and the tendency to spontaneous ossification. Coley (1949) described an atypical subperiosteal giant-cell tumour which had a uniformly good prognosis. Radiotherapy has also given completely satisfactory results, but sometimes the recalcification of the lesion occurred so soon after treatment that it cannot have made any significant contribution to the cure (Brock and Bogart 1945).

It is evident that many aneurysmal bone cysts have an inherent tendency to spontaneous regression, but some become so large that conservative treatment is no longer practicable (Lichtenstein 1953). For this reason they should be treated by excision or curettage as soon as possible. Cysts in the long bones can usually be excised completely, but curettage is the only method for lesions of the neural arch or of the spinous and transverse processes. Operative treatment may be feasible when the cyst is in the body of a thoracic or lumbar vertebra; and even if the curettage is incomplete the results are usually satisfactory. Radiotherapy has the disadvantage of not providing histological confirmation of the diagnosis and should be reserved for inaccessible lesions.

DISCUSSION

Jaffe (1950) was doubtful whether aneurysmal bone cyst was an entity. He suggested that it might follow " a haemorrhagic blow-out in a pre-existing benign bone lesion, in the course of which the original lesion may have been destroyed." There is, however, no evidence in the cases reported here or elsewhere of a parent lesion, and it is unlikely that even a severe haemorrhage could have destroyed all trace of a previous pathological process. Furthermore the common sites for aneurysmal bone cyst are distinctly rare for any other benign lesion of bone. The only feature common to a giant-cell tumour and an aneurysmal bone cyst are the multinucleated giant cells. In all other respects the two conditions are quite distinctive (Table I).

The remarkable feature of an aneurysmal bone cyst is the tendency to heal after incomplete removal or even without treatment, which suggests that it may not be a tumour. Is it then an abnormal response to injury? There is much to support this suggestion. In many of the reported cases, though not in all, the patient's symptoms date from a precise injury. In some a radiograph taken shortly after injury was negative (Bogart and Imler 1947), refuting the assumption that the injury drew attention to a pre-existing lesion. There is a close resemblance between some of the healing cysts and an ossifying haematoma (Cone 1928), and it is possible that the lesion is a peculiar response to a circumscribed subperiosteal haemorrhage. The presence of osteoid tissue and fibre bone in the lesion is an indication of an attempt at repair.

Although there are good reasons for regarding trauma as an etiological factor, it cannot be the only one, for if it were the lesion would certainly be more common.

Since the pathogenesis of the lesion is obscure, the term aneurysmal bone cyst is as satisfactory as any. It is non-committal and describes with reasonable accuracy the pathological features of a well defined disease process which merits individual recognition and treatment.

SUMMARY

1. The clinical, radiographic and pathological features of aneurysmal bone cyst are described and illustrated by case reports.
2. Reasons are given for accepting the lesion as a clinical and pathological entity.
3. The cyst has a tendency to spontaneous regression and healing may occur after partial excision.

My thanks are due to those surgeons who have given me the opportunity of examining the case records of patients under their care, and to Mr Gabriel Donald and Miss Dorothy Davidson for the illustrations. I am especially indebted to Dr M. E. Catto and Dr W. A. Taylor of the Pathology Department of Glasgow University. They first drew my attention to the lesion and were also responsible for the histological preparations.

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ENGELMANN'S DISEASE

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It is appropriate that a publication in honour of Sir Thomas Fairbank should include an account of Engelmann's disease. Not only did he record one of the few cases of this rare disorder, but he saw the first case ever reported (Case 1, Table I) and was responsible, thirty years later (Fairbank 1951) for establishing the diagnosis in this previously undiagnosed case.

The first paper drawing attention to what is now known as Engelmann's disease was published by Camurati in 1922, seven years before Engelmann's account appeared. Previously there had been only the case report (Cockayne 1920) of the then undiagnosed "case for diagnosis" which was recognised later by Fairbank. The literature now contains accounts of a total of twenty-one cases, sixteen of which are reported in detail. This paper adds a twenty-second, a seventeenth detailed report. The five more briefly reported cases are described too scantily for detailed study, but adequately for one to be sure of the diagnosis. These twenty-two cases are all summarised in Table I.

The literature of any pathological rarity must contain accounts of cases in which the diagnosis is open to doubt. It therefore follows that I have rejected some reported cases which other writers have seen fit to accept.* This seems inevitable.

CASE REPORT

The patient was a Welsh woman, twenty-nine years old when I first saw her in 1948. Her family history contained nothing unusual. She had been born at full term, but was said to have been a very small baby. She did not walk till she was three years old, and she was "always weak." She had an attack of jaundice at the age of twelve, and her tonsils were removed when she was fourteen. She was never strong enough to take part in organised games at school.

She had complained as long as she could remember of pains in her knees when she walked. There had been no other symptoms till adolescence, when menstruation did not begin and signs of puberty did not develop. When she was about twenty she began to complain of pain in the back and of constant pain in the legs.

She had her first menstrual period when she was twenty-three years old. For the first six months the periods were regular and normal. They then recurred at decreasing intervals and with increasing loss. After twelve months she was bleeding so severely and continuously that a course of x-ray treatment was given. This stopped menstruation for six months, but the continuous severe bleeding soon recurred. At the age of twenty-five a permanent menopause was produced by a second course of x-rays.

Her other symptoms then increased, and she became unable to stand for more than twenty minutes at a time or to walk more than half a mile (0·8 kilometre).

On examination, she was short (5 feet 1½ inches: 156 centimetres), thin (82 lb.: 37·3 kilograms) and pale. Her skin was normal, though she had very little subcutaneous fat. She stood with a slight tilt of her whole trunk to the right (Fig. 1). Her gait was short-stepping and painful. Trendelenburg's test was negative. Her lumbar spine was almost completely stiff and abduction of the hips was limited to 30 degrees on each side. The other joints all moved

* The case reported by Hirsch (1929), though accepted by Michaelis (1949), is among those I reject. Neither can I accept Holt and McIntosh's (1940) case, even though Fairbank (1951) thinks it "might be included in the Engelmann group." Ribbing in 1949 reported a series of cases of what must be an allied, asymmetrical familial disorder. Neither he nor Garland (1946), nor Smyth, Potter and Silverman (1946), considered the patients they described to be suffering from Engelmann's disease, but later reporters have thought them all "possible examples." I am satisfied that all these reports represent other diseases.

fully. The lumbar spine and all the major long bones appeared very tender, though none of the latter was palpably thickened.

Her muscular development was very poor and her muscle tone so poor that she could put her hands on the ground with her knees straight and with her lumbar spine held stiffly in lordosis.

Secondary sexual characteristics were quite well developed. The lower border of her liver was just palpable and was a little tender. There were small, tender palpable lymph nodes in both groins.

The resting pulse-rate was 90 per minute and her blood pressure 135/90. The heart appeared normal.

There was no discoverable abnormality in the central nervous system, the visual fields, the ocular fundi or the retinal vessels. She appeared normally intelligent.

FIG. 1
Engelmann's disease in a woman twenty-nine years old. (Case 17, Table I.)

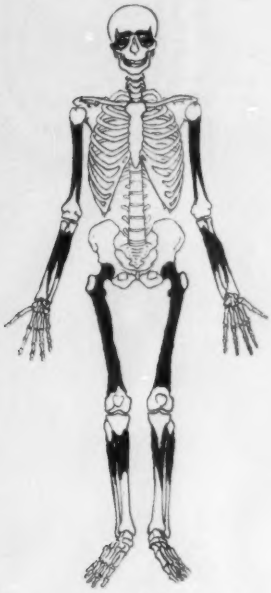

FIG. 2
The distribution of the bony changes in Case 17.

Radiographic examination—There were striking alterations in the radiographic appearances of the skull and of all the major long bones. The distribution of the changes is shown in Figure 2. The skull showed an irregular but symmetrical increase in the density of its base, affecting particularly the sphenoids, the petrous portions of the temporals and the basi-occiput (Figs. 3 and 4). The vault, the facial bones and the jaws were not apparently affected, and the cranial sinuses were all patent. The sella turcica was normal.

The long bones showed a remarkably symmetrical sclerosis of their diaphyses. Subperiosteal and particularly endosteal deposition of a new bone of amorphous appearance produced much cortical thickening. The epiphyses outlined normally, and the metaphyses were clear in all bones except the humeri (Figs. 5 and 6) and the femora (Figs. 9 and 10). The areas of sclerosis in the shafts were fairly sharply demarcated from the normal bone of the ends. The outlines of the medullary cavities were always recognisable, though narrowed. Small erosion pits were visible in many of the sclerosed areas.

Broadening of the affected bones was neither uniform nor striking, and the ends were not " clubbed." There was a valgus bow in the humeri (Figs. 5 and 6), but other bones were not bowed. The forearm bones were more broadened (Figs. 7 and 8) than the humeri; their metaphyses and the lower fourth of each ulna were normal. The sclerosis " flowed " along the interosseous borders of the radii.

The femora were severely affected (Figs. 9 and 10). Only the greater trochanters and the extreme lower ends were free from change. Such involvement of the femoral heads and necks has not been reported in any other case. In this woman it may be associated with her adult age or with her previous pelvic irradiation, or with both.

The tibiae and fibulae (Fig. 11) were affected only in their upper halves. The involved

FIG. 3
The skull: norma frontalis.

portions of the fibulae were as severely affected as any bone, with marked broadening and petrosis and with very clear erosion pits.

The bones of the spine and pelvis, the ribs, scapulae, clavicles, patellae, hands and feet were normal.

Laboratory investigations—The blood and the urine were examined on many occasions. No significant abnormality was ever found in either except glycosuria in one specimen. The fasting blood-sugar never rose above 105 milligrams per 100 millilitres, and glucose-tolerance tests were normal. The blood-urea was never above 28 milligrams per 100 millilitres.

The blood Wassermann reaction was repeatedly negative. The serum alkaline phosphatase varied from 13 to 29 units per cent, calcium from 9·3 to 11·4 milligrams per 100 millilitres,

inorganic phosphorus from 3·0 to 3·2, blood cholesterol from 176 to 188. The serum albumen was 4·7 per cent, globulin 3·3 per cent. The serum carotenes were 70 milligrams per 100 millilitres and serum vitamin A 115 I. U. per 100 millilitres. The 17-ketosteroids were 6·5 milligrams in 24 hours, gonadotrophins 25 m. u. The daily excretion of calcium in the urine varied from 160 to 220 milligrams.

The red-cell sedimentation rate (Wintrobe) was always in the neighbourhood of 20 millimetres in the first hour. The basal metabolic rate was constantly increased, being 25 to 35 per cent above normal on each test.

The sternal marrow was examined by Dr M. C. G. Israëls, who considered it normoblastic, fairly cellular and normally supplied with active granulocytes.

FIG. 4
The skull: norma lateralis.

Biopsy—The upper third of the right fibula was resected (8.11.1948). The excised bone was thick, rough and hard. All tissues around the periosteum were surprisingly fatty.

The biopsy showed osteosclerosis with no specific features (Figs. 12 to 14). The sclerosis was produced chiefly by deposition of mature lamellar bone in the cancellous tissue, producing a much thickened cortex. Abundant endosteal osteoblasts were visible, but an osteoid margin could not be seen. Some of the thickened areas showed osteoclastic erosion with the production of new cancellous spaces. Under the periosteum there was a little new woven bone, but there was not enough of this to suggest rapid new bone proliferation. Specially stained sections showed no sign of lipoid deposition. The marrow appeared normal. There was a little congestion of the vessels and some increase in connective tissue in the spaces in the thickened areas.

FIG. 5
The right humerus.

FIG. 6
The left humerus.

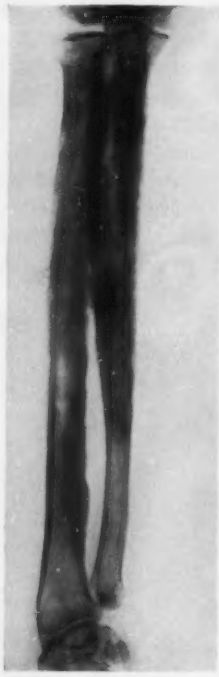

FIG. 7
The right forearm bones.

FIG. 8
The left forearm bones.

FIG. 9
The pelvis and the upper ends of the femora.

Treatment—In view of the menstrual history, dienoestrol (0·6 milligrams per day), later changed to ethinyl oestradiol (0·5 milligrams per day), was tried. The patient gained weight under this treatment, eventually reaching 86 lb. 9 ounces (39·3 kilograms), but she made no other improvement. The usual vaginal bleeding followed withdrawal of the oestrogen.

There seemed to be no other reasonable line to follow. An attempt to relieve her with methyl testosterone (50 milligrams on alternate days) also failed. She is at present using large doses of aspirin, and nothing else.

Progress—In the six years since her investigation her symptoms have become worse. She now has continuous, widespread pain and is confined to her house.

DISCUSSION

This disease is probably less rare than has been thought. Sixteen of the twenty-two accepted cases (Table I) have come to light in the last twelve years.*

The syndrome usually consists of physical under-development, an abnormal gait, and a series of symmetrical hyperplastic bone changes affecting the shafts of the major long bones and, usually, the base of the skull. The hyperostosis appears to be progressive and is likely to reach ultimately the widespread distribution seen in the case reported here. It is clearly difficult to give such a disorder a useful and appropriate name. The accepted eponym overlooks the fact that Camurati's (1922) description preceded Engelmann's by seven years. Titles which refer only to the skeletal changes ignore the fact that this is a general constitutional disorder. " Hereditary symmetrical osteitis of the lower limbs " (Camurati 1922), " Osteopathia hyperostotica (sclerotisans) multiplex infantilis " (Engelmann 1929) and " Infantile poliostotic sclerosis " (Casuccio 1949) all make incorrect assumptions. Probably the best suggestions so far are " Progressive diaphysial dysplasia " (Neuhauser *et al.* 1948) and " Progressive diaphysial hyperostosis " (Gulledge and White 1951), although these names are based solely on the bony changes. At present the name " Engelmann's Disease " has become widely accepted and is understood to mean this disorder and no other.

CLINICAL FEATURES

As shown in Table I, the sex incidence is approximately equal, twelve males to ten females. One patient† (15) was a Filipino and another (16) was Jewish. No case has yet been reported in a Negro or in a patient from the Asian mainland. There has been more than one case in the same family in two Italian reports (Camurati 1922 and Ortolani and Castagnari 1953), and there was a doubt about a brother in one other case (11) (Michaelis 1949).

The youngest age at which the diagnosis has been made is thirty-three months (21) and the oldest fifty-five years (3). Fifteen of the patients reported were children under fifteen years old, six were adults of nineteen or over. Whenever recorded, birth was normal, at full term after normal pregnancy. One patient (15) was a vegetarian; the others had no recorded unusual dietetic habits. One adult patient (13) gave a previous history of pulmonary tuberculosis, otherwise no intercurrent disease was reported. With one exception (12) the children had walked rather late (eighteen months to three years) and dentition had tended to be retarded. In all who were old enough, puberty had been late, the genitalia were never very large and secondary sex characteristics were seldom well developed, though one patient (11), a woman of twenty-four, had a moustache. One male adult (13) is reported to have been sexually normal.

Symptoms always started early in life, nearly always before the age of seven and often

* While this paper was being prepared the author learned of another apparently certain case in a child of three (Bonar Mason 1955).

† Throughout this discussion, numbers in brackets refer to the numbers of the cases in Table I.

TABLE I

ENGELMANN'S DISEASE

a) Summary of seventeen cases fully reported

Case number	Reporter	Age	Sex	Family history	Symptoms	General physique	Muscular development	Deformities
1	Cockayne 1920, Fairbank 1951	9½	M	Negative	Aching in legs, loss of weight, incontinence	Poor	Not commented upon	Flat feet
2 (son of Case 3)	Camurati 1922	7	M	Father also affected (Case 3), possibly several other relatives	Pain in thighs and legs	Poor	Poor	None reported
3 (father of Case 2)	Camurati 1922	55	M	See above (Case 2)	None. Had had pains in legs before adolescence	Poor	Poor	None reported
4	Engelmann 1929	8	M	Negative	Pain in legs since age four	Poor	Poor	Bow legs, lordosis, flat feet
5	Ortolani and Castagnari 1933	6	F	Brother affected (Case 22)	Easy fatigue and poor general health since age two. Pains in limbs	Below normal	Markedly impaired	Lordosis
6	Riley and Shwachman 1943 Neuhauser *et al.* 1948	4¾	F	Negative	Faulty gait, easy fatigue, failure to gain weight	Thin, under weight	Weak	
7	Riley and Shwachman 1943 Neuhauser *et al.* 1948	5	M	Negative	Abnormal gait, tired easily	Under weight		
8	Sear 1948	10	M	Negative	Difficulty in walking since age three		Markedly impaired	Pes cavus, knock knees
9	Neuhauser *et al.* 1948	10	F	Negative	Abnormal gait since age six, failure to gain weight		Weak	
10	Neuhauser *et al.* 1948	4½	F	Negative	Vague pains in limbs		Weak	Flat feet
11	Michaelis 1949	24	F	Negative	Ache in legs since age three		Weak	Lordosis, knock knees, valgus ankles
12	Bingold 1950	14	M	Negative	Poor appetite, retarded growth, vague pain in left knee	Poor	Weak	Ext. rotation deformity of tibiae and valgus feet
13	Stronge and McDowell 1950	28	M	Negative	Pain in legs		Weak	Lordosis, tibial bowing
14	Fairbank 1951	8	M	Negative	Peculiar gait, general weakness		Muscles small but fairly strong	Bow legs, flat feet, thoracic kyphosis

TABLE I—*continued*

Gait	Affected bones	Other findings	Follow-up	Biopsy	Comments
Not commented upon	Skull, humeri, radii, ulnae, femora, tibiae, one fibula				
Broad based, antalgic	Femora, tibiae				
Broad based	Femora, tibiae, fibulae				X-rays quite typical despite unusual history, etc.
Wobbly	Skull, all major long bones	General enlargement of lymph nodes			
Swaying " like a goose "	Tibiae, fibulae (1933), all major long bones (1953)	Palpable spleen	20 years		
Waddling, broad based	All major long bones	Hyperactive deep reflexes, occasional ankle clonus	6 years	Sclerosis	
Straight-kneed, slapping, broad based	Skull, all long bones	Hyperactive reflexes and unsustained ankle clonus	6 years	Sclerosis	
	Skull, all long bones, atlas, ilia, ischia, scapulae	K.J.s and one A.J. absent, spleen and liver palpable		" Nothing abnormal "	
Waddling	All long bones		9 years		Puberty at fourteen, dysmennorhoea and oligomennorhoea
Swaying	Skull, all long bones			Sclerosis, diffuse periosteal thickening	
Uncertain	Skull, all long bones				
Waddling	Femora, tibiae			Endosteal sclerosis the most prominent feature	
Waddling	Skull, all long bones		9 years	" Normal structure "	Thickening and sclerosis progressed long after epiphyses were united
Lurching	Femora only				

TABLE I—*continued*

ENGELMANN'S DISEASE

a) Summary of seventeen cases fully reported

Case number	Reporter	Age	Sex	Family history	Symptoms	General physique	Muscular development	Deformities
15	Gulledge and White 1951	9	M	Negative	Pain and weakness in legs	Poor	Under-developed	
16	Lavine and Koven 1952	12	M		Tiredness on walking	Malnourished	Poor	Flat feet
17	Griffiths (present case)	29	F	Negative	Weakness, pain in legs and back, abnormal puberty	Poor	Poor	Scoliosis, valgus ankles

b) Summary of five probable cases not reported in full detail

Case number	Reporter	Age	Sex	Family history	Symptoms	General physique	Deformities
18	Fritsch 1922	31	F	Negative	Pain in right leg		
19	Casuccio 1949						
20	Geschickter and Copeland 1949	7	M				
21	Gillespie and Mussey 1951	3	F	Negative	Vague leg pains	Well nourished	Lordosis
22	Ortolani and Castagnari 1953	19	M	Sister certainly affected (Case 5)	None		

as soon as walking started. Failure to thrive and to gain weight in early childhood was a frequent presenting symptom, as were easy fatigue and the abnormal gait.

All but seven of the patients complained of pain. Five (6, 7, 8, 9, 16) of those who were free from pain had widespread involvement of the skeleton, but none of these five were over the age of twelve. One patient (3) aged fifty-five said that his pain had disappeared at adolescence. There was no other example of improvement in symptoms.

PHYSICAL SIGNS

Only one patient (13) was said to be tall. Five others (4, 5, 11, 12, 17) were well below average height. The limbs, especially the legs, tended to appear long and slender in proportion to the height.

The general physique was considered poor in all but two cases (7, 21), and no patient over the age of five was regarded as well developed or well nourished. An excessive lumbar lordosis was reported in four (4, 5, 11, 13) and various other postural defects in three (12, 14, 17).

In no case has the gait been noted to be normal. Though usually described as a wide-based waddle, it has in some cases been determined more by pain. Sometimes there was a history of a normal gait until the age of four or five. In no case was Trendelenburg's test reported to be positive.

TABLE I—*continued*

Gait	Affected bones	Other findings	Follow-up	Biopsy	Comments
Wide waddle	Skull, all long bones		2 years	Hypertrophied trabeculae, thickened cortex and periosteum	Progression of disease seen in x-rays over two years
Waddle	Skull, all long bones	Hyperactive deep reflexes		Dense hyperostosis with sudden demarcation	
Coxalgic	Skull, humeri, radii, ulnae, femora, tibiae, fibulae	B.M.R. raised	6 years	Endosteal and subperiosteal hyperostosis	Menarche at twenty-three, artificial menopause at twenty-five

Gait	Affected bones	Other findings	Follow-up	Comments
	Skull, humeri, femora, tibiae. (X-rays not typical)	B.M.R. plus 30 per cent, diminished visual fields		
	Femora, tibiae, fibulae	Anaemia		
	Humeri, radii, ulnae, skull			
Wide-based waddle	Skull, clavicles, all long bones	Umbilical hernia	20 months	
Had been disturbed earlier in life	Femora, tibiae, fibulae (left more than right)		19 years	No change observed in prolonged follow-up

The spleen (5) and the liver (17) were each reported palpable once only. There was no other suggestion of any visceral abnormality.

Swollen long bones were palpable in the thin lower limbs of four cases (4, 5, 7, 8) and in the upper limbs of one (4). Joint movement was full in all except five cases (4, 5, 14, 17, 21), in three of which (5, 17, 21) there was appreciable limitation of hip movement, in one (4) limited extension of the knees, and in one (14) limitation of both hips and knees. The muscular hypotonus produced excessive straight-leg raising in three (2, 3, 17).

Muscular weakness and under-development was reported in all fully described cases except three (7, 14, 16), two of which were malnourished or under weight, while the third (14) had muscles which were " small but fairly strong."

In six cases (6, 7, 8, 9, 14, 16) the reporters considered that there were significant signs of disturbance of function in the central nervous system. These signs were always somewhat equivocal, increased tendon reflexes in three (4, 6, 16), decreased or absent in three (8, 9, 14), inconstant ankle clonus once (7), never an extensor plantar response or any sensory change.

THE BONY LESIONS

The changes shown in the bones have never differed in quality from those described in the case reported here. The lesions have always been restricted to the diaphyses in the long bones, and usually to the middle two-fourths of them. Involvement of any metaphysis or

epiphysis has not previously been reported. In children, especially in the femora, a patch of relatively normal shaft has sometimes been seen between one small area of sclerosis in the upper part of the diaphysis and a much larger area lower down (5, 6, 8, 19). All the bony changes have been strictly symmetrical except for one instance (1) in which one fibula was unaffected.

There were changes in the skull in all except eight cases (2, 3, 6, 9, 12, 14, 19, 22), of which two were in adults (3, 22). The changes resembled those seen in the case reported in this paper (17), except that the vault as well as the base was thickened in five cases (5, 8, 11,

FIG. 10
The lower parts of the femora.

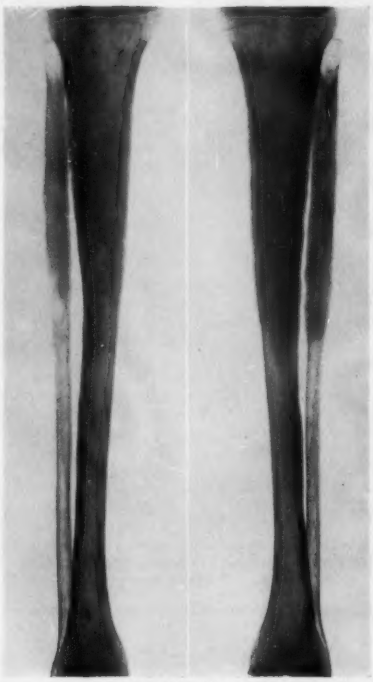

FIG. 11
The tibiae and fibulae.

13, 18). The vault was mainly affected in the frontal bones. Changes have never been reported in the parietals or in the squamous parts of the temporals, and the facial bones and jaws have not been affected. The sella turcica was slightly expanded in one case (18), in which there was " some restriction " in the visual fields.

There is only one report of involvement of any part of the spine (8), and then only the atlas was affected. The ribs became involved in only one case (5) and the pelvis in another (8), which latter was the only example of changes in the scapulae. The middle thirds of the clavicles were affected in three cases (7, 8, 10), the inner thirds in a fourth (5), and in one more (21) involvement of the clavicles is reported without description of the pattern. The humeri were affected in all cases except six (2, 3, 12, 14, 19, 22) and involvement of the forearm bones is not reported in those six or in two others (1, 18).

The most striking changes have been those seen in the femora, which have never been reported to be normal, though involvement of those bones was not noticed in one case (18) until the patient was twenty-nine years old. The tibiae were stated to be affected in all but two (14, 20) of the reported cases, though their involvement was always less marked than

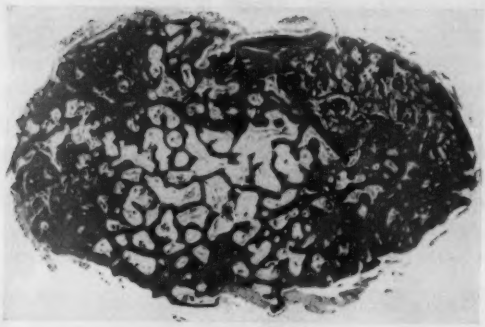

FIG. 12

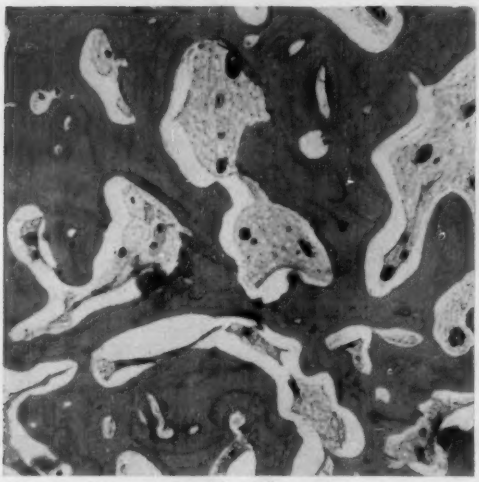

FIG. 13

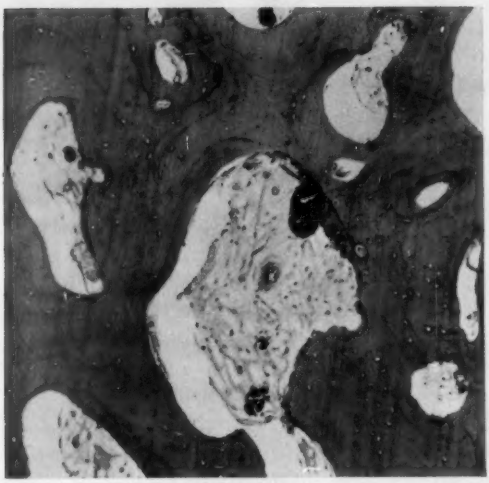

FIG. 14

Histology of the upper end of the shaft of the right fibula: Figure 12 = ×5; Figure 13 = ×40; and Figure 14 = ×70. Note the cortical thickening, the regular lamellar structure of the bone, the abundance of osteoblasts and the absence of any abnormal cell or tissue.

that of the femora. If the tibiae were affected, so were the fibulae in all but another two cases (12, 18) and in one case (1) where only one fibula was abnormal.

There are no reports of involvement of any other bones.

LABORATORY INVESTIGATIONS

The blood showed a slight monocytosis in two cases (4, 11) and a lymphocytosis of 40·5 per cent with a 5 per cent eosinophilia in another (4). Otherwise the count has never shown more than a mild anaemia. There have been no significant biochemical changes reported, except a raised basal metabolic rate in two adults (17 and 18).

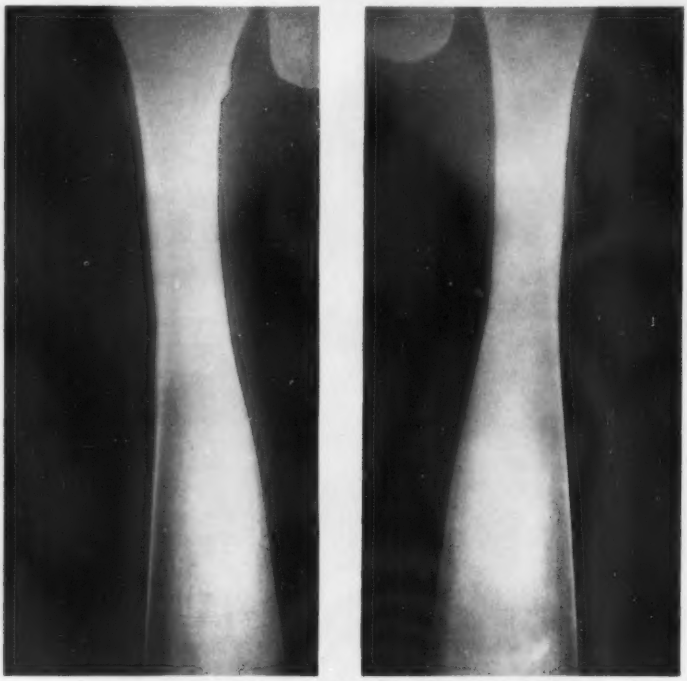

FIG. 15

Femora in a case of familial metaphysial dysplasia. Note the absence of sclerosis in the affected bones. (Films of a patient of Dr E. B. Sundaram, reproduced here by his kind permission.)

HISTOLOGY

Biopsy has been performed in seven children (6, 7, 8, 10, 12, 15, 16) and in two adults (13, 17). Some of the reports are very brief, but it is clear that the microscope has shown osteosclerosis every time, and little else. No tissue has ever been seen which was not of the structure of normal bone. There has never been any suggestion of inflammatory change.

DIFFERENTIAL DIAGNOSIS

The diagnosis of Engelmann's disease is usually quite obvious. It is, indeed, difficult to justify the attempts made to suggest that a differential diagnosis exists, and the fact that previous reporters have between them suggested that twenty-five other diseases are worthy of consideration in this respect is more a testimony to their ingenuity than to the realities of the problem. A little consideration will exclude nearly all of them.

In a disease so rare as this, the chief diagnostic problem is less one of exclusion than of inclusion. The difficulties lie in the " border-line " cases, such as that of Garland (1946), in which symmetrical sclerosis of the major long bones of the appendicular skeleton accompanied a variety of leontiasis ossea. There was, however, no general disturbance in this patient, the metacarpals were affected, and the pattern of the bone change did not resemble that of Engelmann's disease.

In young children there may be justifiable difficulty in distinguishing Engelmann's disease from infantile cortical hyperostosis (Caffey and Silverman 1945). This latter disease, however, has an acute febrile onset in early infancy, the sclerosis is not symmetrical, the mandible is always involved, and the affected bones become normal in about two years.

FIG. 16

FIG. 17

Right and left tibiae from the same case of familial metaphysial dysplasia.

Familial metaphysial dysplasia (Pyle 1931, Bakwin and Krida 1937) produces symmetrical changes in long bones, which become broadened and clubbed (Figs. 15 to 17). This venerable disease, which was the probable cause of the changes found in a femur in an ancient Nubian cemetery (Smith and Jones 1910), produces no true sclerosis though it has a superficial resemblance to Engelmann's disease in other respects.

THE NATURE OF THE DISEASE

It is interesting but futile to speculate upon the essential pathology of Engelmann's disease. Almost nothing is known of the systemic changes. There has not yet been one case examined post-mortem. The early onset, the absence of abnormal cell-structure in such tissues as have been examined and the symmetrical progress in the skeleton all suggest a genetic origin. No known toxin, no deficiency and no endocrine imbalance alone produces anything like this.

I wish to thank Professor S. L. Baker for help with histology, Dr M. C. G. Israëls for examining my patient's sternal marrow, and Dr Raymond Whitehead for help with Italian literature. The photomicrographs are the work of Mr T. C. Dodds of Edinburgh.

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POLYARTERITIS NODOSA WITH NEW BONE FORMATION

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Polyarteritis nodosa was first described as a clinico-pathological entity by Kussmaul and Maier (1866), who called it periarteritis nodosa. Ferrari (1903) first called the disease polyarteritis nodosa, a name that describes the pathology more accurately.

The disease may be grouped with the collagen diseases—desseminated lupus erythematosis, scleroderma, rheumatoid arthritis and others. It is characterised by patches of acute inflammation, necrosis of cells and fibrinoid changes in the collagen fibres of small and medium-sized arteries. The symptoms depend either on ischaemic changes in the tissues supplied by the affected vessels or to rupture of vessels. It is not surprising, therefore, that symptoms referable to practically every system and organ have been described alone or in combination. It is the presentation of symptoms from disease processes involving many systems in a bizarre fashion that usually suggests the possibility of the diagnosis. Arthralgia is common but arthritis rare (Lowman 1952). The medical literature is well reviewed in English by Curtis and Coffey (1934), Harris, Lynch and O'Hare (1939), Miller and Daley (1946) and Nuzum and Nuzum (1954). None refers to affection of bone in this disease. The opportunity is taken of reporting a case of polyarteritis nodosa, the presenting and only symptoms of which were pain and swelling of the shins with subperiosteal new bone formation. Investigations including biopsy are recorded, as well as the response to treatment.

CASE REPORT

A man of fifty-eight, a bank messenger, had been perfectly fit until November 1953, when he noticed redness of the skin over his right shin. At first this was painless and seemed to subside after ten days. Four months later swelling became localised to the middle third of the right tibia and a similar condition appeared in the left leg. He now had severe throbbing

TABLE I

ROUTINE BLOOD EXAMINATION

Date	Haemoglobin (*per cent*)	P.C.V.	E.S.R.		W.B.C.
3.54	108	50	19		7,200
4.54	—	43	30		7,000
3.7.54	101	41	34		8,000
10.7.54	—	38	44		8,500
8.54	—	45	20	Cortisone therapy	
9.54	74	43	15		
10.54	103	45	20		
11.54	101	46	23		
6.55	—	—	14		

pain in the lower part of both legs which kept him awake at night. It was aggravated by standing or walking, so that he was unable to work or to get about the house. Otherwise, his general health was good.

There was no significant family history except that one brother had died of diabetes mellitus. He had had no serious illness in his life but had travelled throughout the Far East during the 1914–1918 war.

Examination revealed a heavily built plethoric middle-aged man. The blood pressure was 190/120. The heart was not clinically enlarged. Fundi showed Grade I hypertensive changes. Pitting oedema was present over the dorsum of both feet. There was some swelling, redness and heat over the subcutaneous border of the tibiae, over the upper two-thirds, worse on the right. This area was tender to pressure. The skin was adherent to bone and was desquamating on the right. There were no other abnormal signs in any system.

Radiographs—No active lesion was seen in the lung fields. The affected bones are shown in Figures 1 to 5.

FIG. 1 FIG. 2 FIG. 3 FIG. 4

Figure 1—Radiograph of left tibia and fibula taken immediately after biopsy. Note subperiosteal new bone round fibula and commencing along interosseous border of tibia. Figure 2—Right tibia and fibula. Initial radiograph March 1954. Note subperiosteal new bone on medial and lateral aspects of upper third of tibia and also round shaft of fibula. Figure 3—Right tibia and fibula three months later. Note great increase in subpericsteal new bone in upper third of tibia, and commencing new bone formation on medial surface of lower third of tibia. Figure 4—Right tibia and fibula a year later. The newly formed bone has increased and consolidated.

Pathological investigations—The blood Wassermann and Kahn reactions were repeatedly negative. Agglutinations against the salmonella group and brucella group of organisms were repeatedly negative. The results of routine blood examinations are shown in Table I.

Serum calcium, phosphate and alkaline phosphatase were all normal. Blood urea 24 milligrams/100 millilitres. Urea clearance 63 per cent of normal. Serum ascorbic acid 0·6 milligrams/100 millilitres. After 800 milligrams of ascorbic acid by mouth, serum levels rose to 1·7 milligrams/100 millilitres. It was concluded that he did not suffer from scurvy.

The *urine* repeatedly showed a trace of albumen and an occasional red cell and pus cell.

Biopsy—A segment of fibula with adjacent soft tissue was taken. Macroscopically the subcutaneous tissue appeared oedematous and the muscle was pale and contained small gelatinous areas. The histology showed polyarteritis. The biopsied muscle showed atrophy of fibres and diffuse fibrosis (Fig. 6). There was marked patchy infiltration of the tissue with inflammatory cells—chiefly lymphocytes and plasma cells, but also neutrophil and eosinophil polymorphs. Some of the inflammatory cells were situated in the interstitial connective tissue of muscle bundles; many were in the perivascular tissue surrounding small blood vessels (Fig. 7). Occasionally the walls of small arteries were involved by the lesions, all layers containing inflammatory cells and showing varying degrees of destruction (Figs. 8 and 9). A considerable amount of periosteal new bone was present on the surface of the fibula (Figs. 10 and 11). The deeper cortical bone was histologically normal, and neither the newly formed bone nor the adjacent periosteal tissues showed involvement by inflammatory lesions.

FIG. 5
Left foot. Note subperiosteal new bone round the shaft of the first metatarsal.

DISCUSSION

Polyarteritis nodosa may occur at any age, the youngest recorded case being in the first month of life (Wilmer 1945), but in the 175 cases reviewed by Nuzum and Nuzum (1954) the average age was 41·5 years and males predominated over females in the ratio 4:1. The patient whose case is reported in this paper also had hypertension, which probably occurs in more than half the cases and is due to renal involvement, which manifested itself in this case by albuminuria and diminished urea clearance. The unique feature was the new bone formation which was bilateral and presumably due to stimulation of the periosteum by the arteritis in the surrounding muscles. The bone and periosteum were not directly involved.

The etiology of the disease is uncertain. The present trend is to regard it as a hypersensitivity reaction. Cases have been reported after serum sickness (Rich 1942), iodine treatment of goitre (Rich 1945) and organic arsenicals (Miller and Nelson 1945). Asthma was associated with the disease in 18 per cent of the cases reviewed by Wilson and Alexander (1945). Polyarteritis nodosa has been experimentally produced in rabbits by establishment of an anaphylactic state analogous to human serum sickness (Rich and Gregory 1943). The disease, then, seems to be related to allergy—immunity mechanism in general. There was nothing in the history of this patient, however, to suggest an allergic illness, neither had he taken any drugs to which he might have become sensitised.

With all this evidence suggesting that the disease is in the nature of a hypersensitivity reaction, it would seem reasonable to treat it with corticotrophin or cortisone. Most authorities are agreed that the disease is usually fatal, although Miller and Daley (1946) demur in this belief, and suggest that mild chronic and localising cases, such as two they describe, are often missed. Baggenstoss, Shick and Polley (1951) reported two cases treated with cortisone, in which there were remission of symptoms and lowering of the erythrocyte sedimentation rate on administration of the drug, with partial relapse on stopping treatment and further remission on restarting it. Lovell and Rose (1955) reported eight patients observed

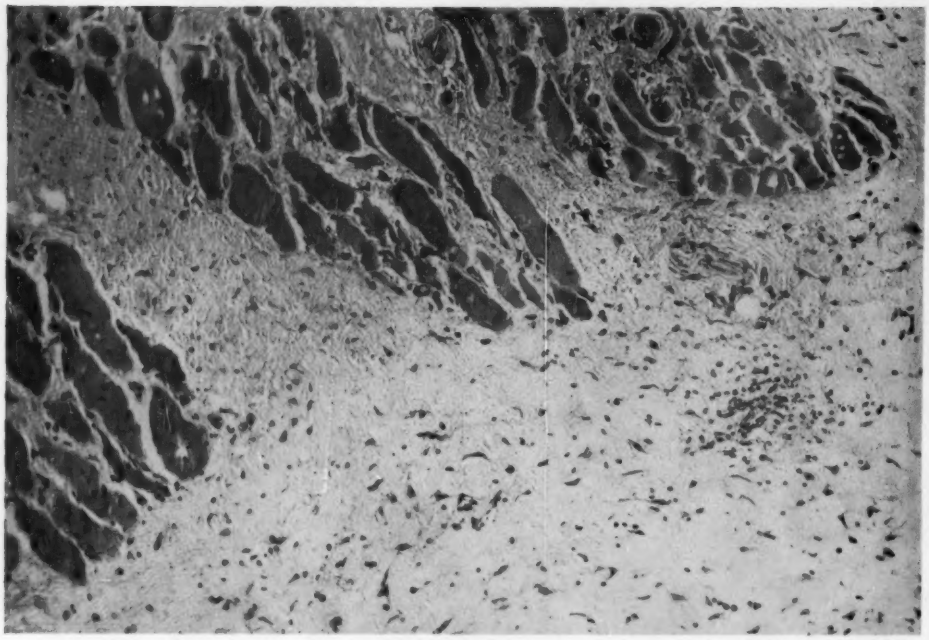

FIG. 6
Muscle atrophy and fibrosis of the affected muscle. (H. and E., ×150.)

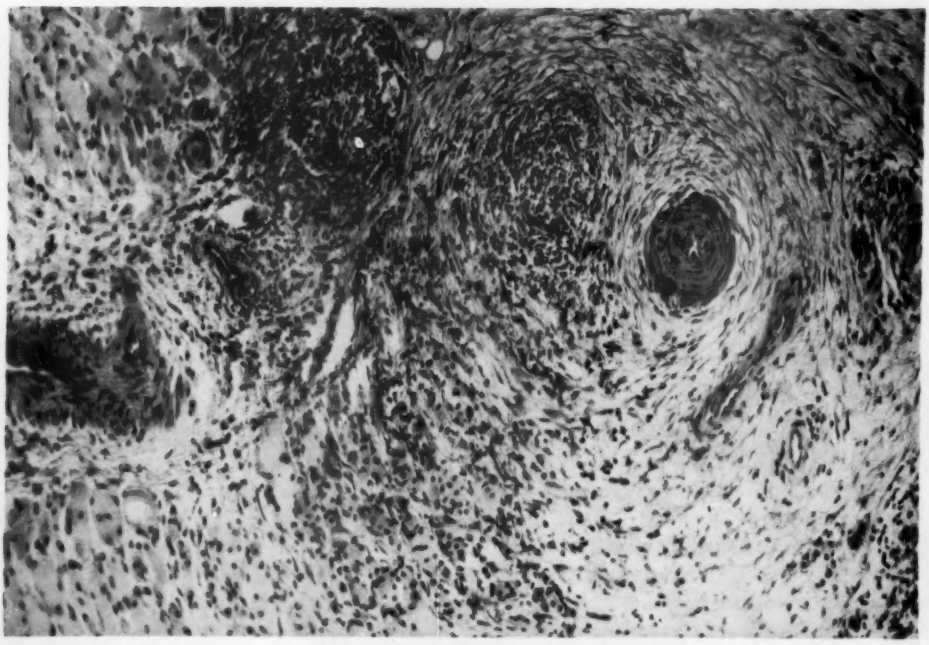

FIG. 7
Muscle. Tissue infiltration with inflammatory cells, showing the perivascular distribution. (H. and E., ×150.)

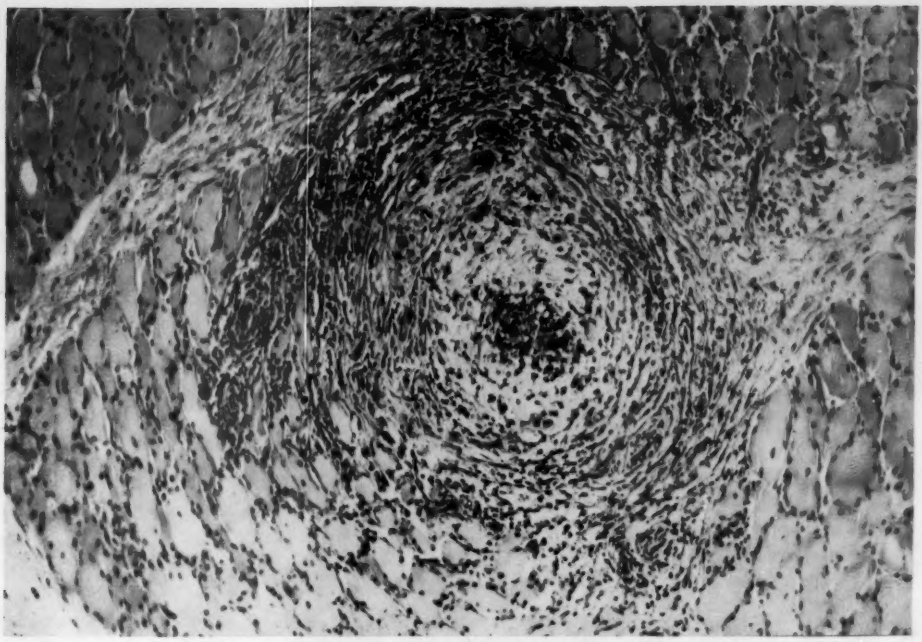

FIG. 8

Muscle. Small artery showing complete disorganisation of wall by inflammatory tissue. (H. and E., ×150.)

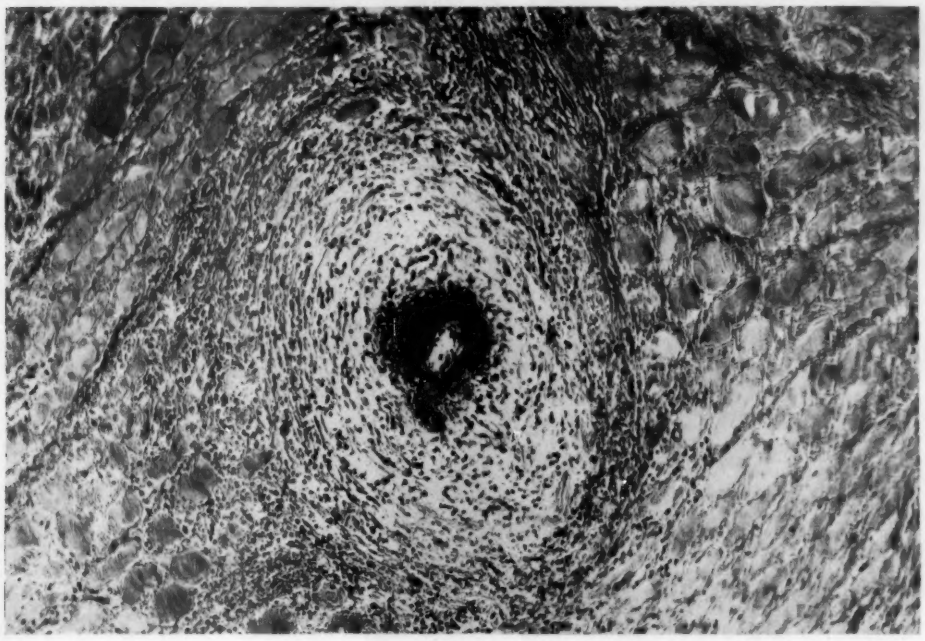

FIG. 9

Muscle. Small artery. The subintimal connective tissue of the vessel wall shows fibrinoid degeneration. (Phospho-tungstic acid haematoxylin, ×150.)

FIG. 10
Fibula. Slab-radiograph of resected tissue, showing conspicuous periosteal new bone. (×4½.)

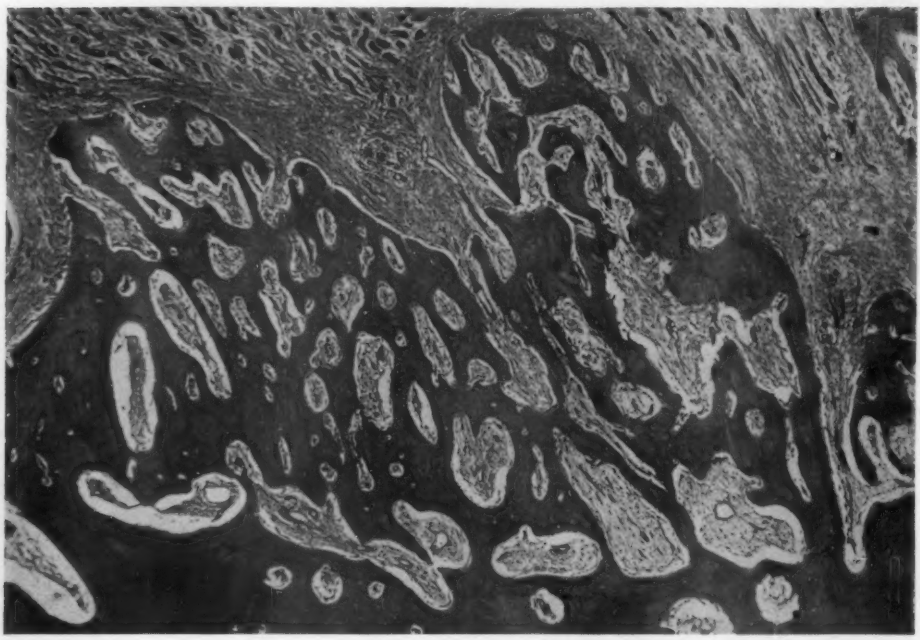

FIG. 11
Fibula. Periosteal new bone, showing the absence of any inflammatory lesions in this tissue. (H. and E., ×35.)

for five years and treated with large doses of cortisone aimed at complete supression of the disease. Two patients died during treatment from malignant hypertension, three patients were in remission and three patients still required cortisone. These authors stress the danger of renal lesions and the importance of early diagnosis and complete suppression of the disease in order to prevent further renal involvement which is irreversible.

The patient whose case is reported in this paper has not had enormous doses of cortisone. His erythrocyte sedimentation rate is down but still slightly above normal. At the same time, on 75–100 milligrams of cortisone daily, the patient's symptoms have resolved completely and he is able to do a full day's work; he has not been troubled by any of the unpleasant side effects of cortisone. Now he feels well and his diastolic blood pressure is about 20 millimetres lower than it was a year ago.

SUMMARY

1. A case of polyarteritis nodosa is reported, the presenting manifestation of which was subperiosteal new bone formation in both legs.
2. The effect of cortisone on the symptoms is recorded.
3. The histology is reported after biopsy of bone and adjacent tissue.
4. Skeletal involvement in polyarteritis nodosa is believed to be hitherto unrecorded.

I wish to thank Mr H. Jackson Burrows for permission to publish this case and Dr H. A. Sissons for the photomicrographs and the histological report.

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CONGENITAL VERTICAL TALUS

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There is a type of congenital flat foot which is clearly associated with a vertical disposition of the talus. It is uncommon and little has been written about it (Hohmann 1934, Niederecker 1955). Two personal observations may give some indication of its comparative rarity. In thirty years of orthopaedic practice I have encountered the deformity in young children only on four occasions, and in a special clinic dealing with congenital foot deformities in the newborn this deformity has been found only once in 131 consecutive cases observed over the past eight years. Of these, 121 were club feet; the remaining ten were flat feet and included a single case of vertical talus. My interest has been directed to the deformity by its unsightliness, by its resistance to conservative and operative treatment, and by the cosmetic and functional blemishes it causes by the time adolescence is reached (Figs. 1*a* and *b*).

DIAGNOSIS

The clinical picture is that of a rigid and flat longitudinal arch with marked prominence of the head of the talus on the medial and plantar aspects of the foot, and eversion of the heel (Fig. 2*a*). There is usually an appearance of dorsal prominence at the tarso-metatarsal junction as if an effort were being made at that level to compensate for the flattening of the hindfoot.

Though these findings should cause suspicion of this deformity in a child, it is the radiograph which is so characteristic (Figs. 2*b* and *c*). The talus lies almost vertically in the line of the long axis of the tibia and the forefoot deviates laterally at the mid-tarsal level. Consequently the head of the talus projects downwards and medially, and only the posterior portion of the superior articular surface of the talus is contained in the tibio-fibular mortise. There is incongruity of the subtalar joint—it looks as if the talus is sliding off the calcaneum, which is more horizontally disposed and more posteriorly placed than normal. Dorsal arching at the tarso-metatarsal level can be seen in the earliest films.

The combined clinical and radiographic appearances give the impression that the hindfoot has broken into two units—the talus on the one hand, and the calcaneum, cuboid, navicular and cuneiforms on the other—and that a subluxation has occurred at the talo-navicular and subtalar joints, the heel and the remainder of the foot being twisted off the talus so as to lie in eversion.

TREATMENT

Until eighteen months ago, when the operation about to be described was devised, I had been unable, like most other surgeons, to control the deformity either by conservative or by operative treatment. In two patients treated just before the outbreak of the second world war, repeated manipulations with and without anaesthesia and prolonged plaster-of-Paris splinting failed to produce the slightest correction. In one an open correction was attempted, the talus being extensively freed on all aspects; in spite of prolonged splinting the deformity relapsed and the situation was made worse by the development of avascular necrosis in the talus. Eventually in adolescence both these children had to have triple arthrodesis, and the patient with the avascular talus, arthrodesis of the ankle joint.

Depressed by these results I was glad to enjoy a lapse of nearly fifteen years before encountering this deformity in a child again. Early in 1953 a boy aged two was seen with typical deformities of both feet which had resisted conservative treatment since birth. Further

FIG. 1*a*

R. K., aged fourteen years, a case of bilateral uncorrected congenital vertical talus. The navicular articulated with the dorsal surface of the neck of the talus and encroached on the articular surface of the ankle joint. The calcaneum was horizontal and the foot was flat. It was also painful.

FIG. 1*b*

A radiograph five years after excision of the navicular and head of talus, and talo-cuneiform fusion using cancellous bone chips. Both feet were now more shapely and painless. (Note dorsal bowing at tarso-metatarsal level.)

manipulations and wedged plasters for another year failed to alter the position of the talus (Fig. 2*d*). The parents, distressed by the appearance of the feet and the clumsiness of his gait, asked that something more should be done. With reluctance and with only a glimmer of a preconceived plan, I operated on the right foot in May 1954 when the boy was three years old.

FIG. 2*a*

N. N., aged two and a half years. Typical deformities are shown, worse on the left side. Note the eversion of the heels, the prominence of the head of the talus on the medial side, and the arching at the tarso-metatarsal junction, especially on the outer aspect of the foot.

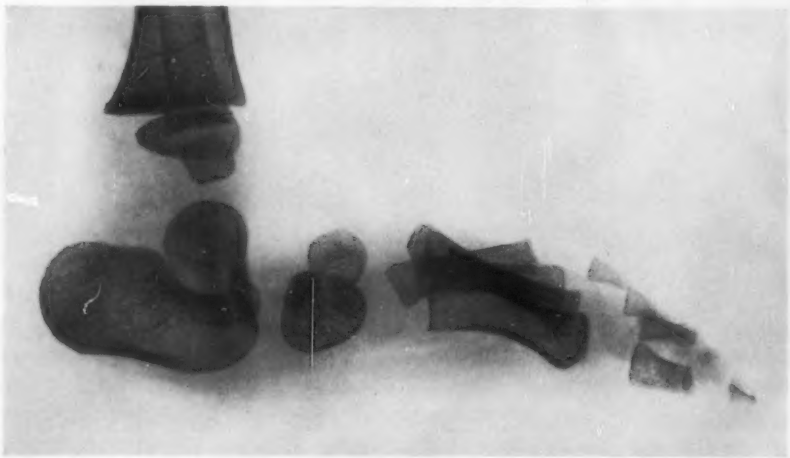

FIG. 2*b*

A lateral view of one foot showing the vertical talus, the disrupted subtalar joint, the horizontal calcaneum, and the dorsal bowing at the tarso-metatarsal junction.

FIG. 2*c*
An antero-posterior view of the same foot. The talus points medially and there is marked outward deviation at the mid-tarsal level.

FIG. 2*d*
A radiograph showing no alteration of the deformity following conservative treatment.

OPERATION

Two incisions were used, but it was decided not to strip the talus widely so as to avoid serious damage to its blood supply. Through the medial incision it was possible to see that the spring ligament was being stretched by the head of the talus, but it was not thin and had no abnormal connections. The tendons of tibialis anterior and posterior seemed normal in appearance and attachment, but to be quite sure of this would of course have required more extensive dissection than was permissible. The articular surface of the head, pointing steeply downwards and inwards, was covered only by capsule and ligament and was not in contact with the articular surface of the navicular.

The lateral incision was made as for subtalar and mid-tarsal fusion. When the interosseous talo-calcaneal ligament, the outer capsule of the subtalar joint and the dorso-lateral capsule of the talo-navicular joint were divided, the navicular was found lying on the dorso-lateral aspect of the neck of the talus. By inserting a skid into the talo-navicular joint the head of the talus could be coaxed dorsally and laterally while the remainder of the foot was manipulated medially until the navicular and calcaneum came into normal contact with the talus. This was done without much difficulty, but on withdrawing the skid the talo-navicular dislocation recurred.

It seemed that something to anchor the talus in a more or less horizontal position was necessary. The procedures advocated by Chambers (1946) and by Grice (1952)—both excellent for mobile flat feet—did not seem applicable in a foot so rigid and in such a young child. It seemed however that the peroneus brevis might act as an anchor and as an active evertor of the talus. Its tendon was detached from the base of the fifth metatarsal, freed as far back as the tip of the lateral malleolus, passed through a vertical tunnel in the neck of the talus and sutured back on to itself (Fig. 3). The talus then remained snugly in normal contact with the navicular with little external support, and the eversion of the calcaneum had been overcome. A plaster-of-Paris cast was maintained for twelve weeks in the over-corrected position.

Eighteen months later the correction of the foot is maintained—indeed it is slightly over-corrected, the heel being rather too inverted and the forefoot too adducted (Fig. 4*a*). There is some irregularity of the neck of the talus due to the tunnel for the tendon and due no doubt also to vascular disturbances, but the body of the talus is normal (Figs. 4*b* and *c*).

Within recent months the same operation has been done on this boy's opposite foot and on two other similar feet. In each case the immediate correction has been gratifying, though it has not been possible always to maintain the talus in its normal horizontal position.

COMMENT

This communication must be regarded as a preliminary report. It is published now without apology for the paucity of material because the condition is so rare and because in my hands the procedure described has been the only effective one. No doubt the operation can be improved by modifications. For example, it might be better to pass the peroneus brevis tendon in front of the lateral malleolus, and it should be possible to avoid over-correction. It will be interesting to watch the development of these four feet and observe what happens to the subtalar and mid-tarsal joints. At this stage it seems fair to say that the foot can be made shapely, though certainly it would be unwise to prophesy about the distant future. But even should painful osteoarthritis develop—and this seems likely in the talo-navicular joint at least—the shapeliness of the foot should make it much easier to give a good cosmetic result by triple arthrodesis than if the deformity had been allowed to persist.

No firm views can be expressed about etiology. Normally the neck of the foetal talus is long and is directed downwards and inwards at an angle of 150 degrees to the axis of its

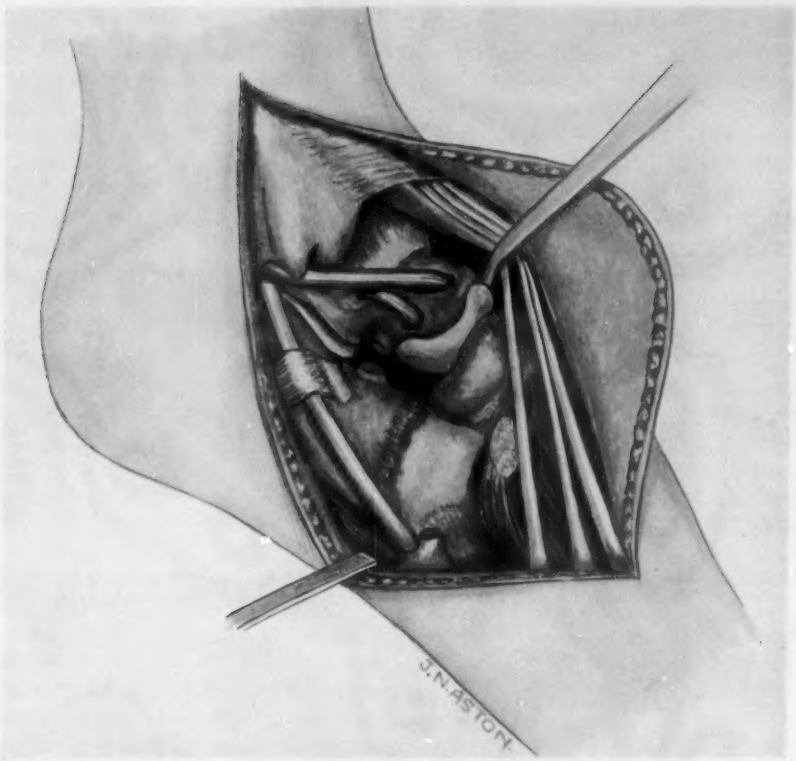

FIG. 3

A drawing of the operation. The interosseous talo-calcaneal ligament, the outer capsule of the talo-calcaneal joint and the dorso-lateral capsule of the talo-navicular joint are divided. The medial talo-navicular ligaments and the capsule of the ankle joint are left intact. The head of the talus is levered outwards and upwards and held there by transplanting the tendon of peroneus brevis through the neck. The lateral displacement of the calcaneum and navicular is reduced.

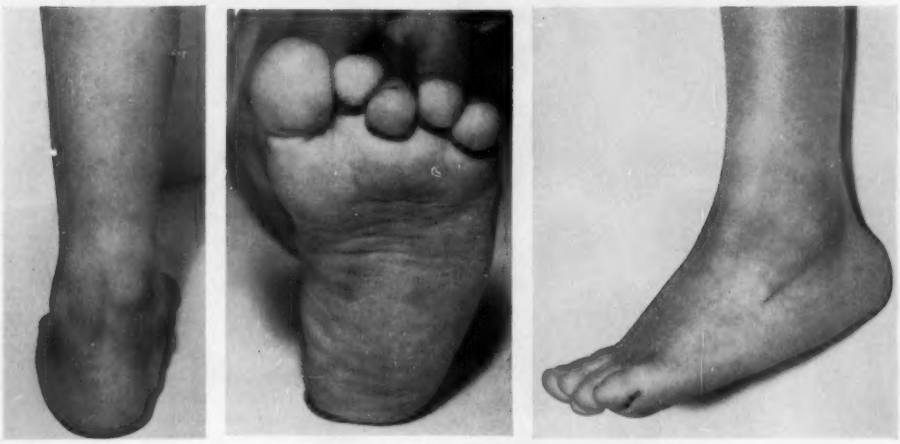

FIG. 4*a*

The same patient, N. N., as was shown in Figure 2. Clinical photographs eighteen months after operation now show a slightly inverted heel, absence of prominence of the head of the talus on the medial border of foot, and loss of the dorsal arching at the talo-metatarsal level.

FIG. 4*b*

A lateral radiograph eighteen months after operation. The position of the talus has been maintained. There is some irregularity of the head and neck due mainly to the tunnel for the tendon.

FIG. 4*c*

The antero-posterior radiograph. The ossific centre for the navicular is now showing clearly. It is lying too far medially, producing a slight varus at the mid-tarsal level.

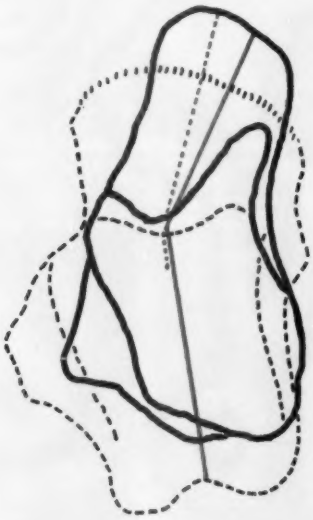

FIG. 5

Outlines of a left talus seen from above (after Keith). This drawing shows the difference between the axis of the body and of the neck of the talus in the foetus (continuous line)—150 degrees, and the adult (dotted line)—160 degrees. Note how the neck is longer and thinner and points more medially in the foetus.

body (Fig. 5); the whole foot is inverted to correspond with this angulation of the neck of the talus. With growth the neck becomes shorter and lies more in the axis of the body, the angle being now about 160 degrees; eversion of the foot occurs at the same time (Keith 1933, Wood Jones 1944). It would appear that at any early stage, perhaps in intra-uterine life, the foot is displaced laterally off the talus. It is probable that this is merely the severest form of congenital talipes valgus; a similar disposition of the talus occurs in extreme degrees of paralytic flat foot and I have observed it in a flat foot in a case of arthrogryposis. The idea that club foot is essentially a talo-navicular subtalar subluxation has always appealed to me; I am inclined to regard "vertical talus" as a subluxation at the same joints but with displacement in the opposite direction. Varying degrees of it can of course occur. In both deformities the rational treatment is to place the navicular and calcaneum in normal relationship to the talus.

It has been observed at operation on older patients that the spring ligament is very thin and it has been suggested that congenital weakness of this ligament allows the head of the talus to displace plantarwards and medially (Fisk 1955). In our last four operations the ligament was carefully examined—hence the medial incision—and appeared to be quite normal, as were the adjacent tendons. If the deformity remains uncorrected it is, of course, easily conceivable that the ligament will gradually stretch and thin out from pressure of the dislocated head of the talus.

SUMMARY

1. Attention is drawn to that type of rigid congenital flat foot in which the talus lies vertically instead of horizontally.

2. This deformity is rare, but if not treated successfully leads to an ugly, painful foot in adolescence.

3. The experience of five patients forms the basis of the present preliminary account. One adolescent and two young children under the age of five were treated unsuccessfully by both conservative and operative measures. More recently in two children with bilateral deformity open operation has been successful in restoring the shape of the foot.

4. The operation is essentially a reduction of a subluxation at the talo-navicular and subtalar joints. It entails freeing the head of the talus sufficiently to allow it to be lifted dorsally and laterally. The talus is then anchored in position by transplanting the distal end of the peroneus brevis tendon through the neck of the bone.

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FEMORAL OSTEOTOMY IN EXTRA-ARTICULAR ARTHRODESIS OF THE TUBERCULOUS HIP

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The usual end-result of uncomplicated tuberculous disease of a joint is fibrous ankylosis. This particularly applies to the hip. With the discovery of antibiotics having a specific action on the tubercle bacillus it was hoped that a rapid and permanent cure of the disease might be obtained. It was also hoped that a mobile joint might be the common end-result and fibrous ankylosis the rarity. So far this desirable state of affairs has not materialised. This is due in part to the inability of the specific drugs to obtain access to the organism on account of the normal local reaction of the body to the disease. It is also often due to delay in diagnosis, so that bone and cartilage damage has occurred before specific drug treatment is started.

There is no doubt, however, that more rapid healing of tuberculous lesions of the hip and knee has been obtained under the influence of streptomycin and other specific drugs. It is also true that a higher percentage of mobile tuberculous joints has followed specific antibiotic treatment (Stevenson 1954). However, it seems probable that some of these mobile joints, on account of intra-articular damage, will give rise to trouble in the future.

For many years the best method of obtaining bony ankylosis of a quiescent tuberculous hip with fibrous ankylosis had been a surgical problem, and it still is. Many methods have been tried, as evidenced by the large amount of literature on the subject. It will be noted that, of the exponents of extra-articular arthrodesis, Brittain was the first to describe, in his ischio-femoral arthrodesis, the simultaneous combination of an extra-articular bone graft with osteotomy of the femur (Brittain 1941, 1942, 1948). Trumble first described the ischio-femoral bone graft but without osteotomy (Trumble 1932, 1937, Van Gorder 1949). Some form of upper femoral osteotomy alone has been used by many surgeons in an attempt to obtain bony ankylosis across the tuberculous hip, but the results have always been uncertain (Wilkinson 1947). Nevertheless the strong fibrous ankylosis often obtained by this procedure can give an excellent functional result in many cases, though it often allows deformity to occur subsequently.

Milch (1947) stated: " in the quiescent phase the likelihood of successful fusion is much greater than in the active phase . . . In this (quiescent) phase the primary effect of arthrodesis may in itself be nothing more than that of enforcing rest upon which the normal processes of repair depend." An intertrochanteric or subtrochanteric osteotomy of the femur alone facilitates this rest, and when combined with an extra-articular bone graft the degree of rest at the damaged joint is even greater. This combination can be carried out simultaneously, as in Brittain's operation, or the osteotomy may be performed a short time after the grafting operation, without interfering with the immobilisation.

Not infrequently an extra-articular bone graft alone results in failure. In many such cases a femoral osteotomy, done primarily to correct deformity at the hip, will produce the desired result of bony ankylosis across the joint with consolidation of the graft (Figs. 1 to 3).

CLINICAL MATERIAL

In this paper I propose to give the results obtained at the Royal National Orthopaedic Hospital of two methods of extra-articular arthrodesis of the quiescent tuberculous hip with

fibrous ankylosis. These two methods are the ilio-femoral graft and the ischio-femoral graft. Between 1933 and 1953, 147 such operations were carried out.

In the ilio-femoral technique the graft is obtained either from the ilium—preferably full thickness—or from the tibia. Frequently such a grafting operation has been followed by an osteotomy of the upper end of the femur. This has been done either within a few months of the grafting operation to correct a pre-existing deformity, or at a considerably later date to correct subsequent deformity which has occurred as a result of failure of the grafting operation.

FIG. 1

FIG. 2

FIG. 3

Case 1—Tuberculosis of the right hip in a boy. Figure 1—Radiograph at age thirteen, showing failed ischio-femoral graft (posterior approach). Figure 2—Three months after subtrochanteric osteotomy there is union of the graft. Figure 3—Eight months after osteotomy there is bony fusion across the joint.

In the ischio-femoral method the graft has been combined as a routine with an osteotomy of the femur as described by Brittain and later modified by Foley (Foley 1949, Cholmeley and Nangle 1951).

Apart from the type of arthrodesis there are certain other factors to consider, such as the age of the patient at the time of operation. It has been found that the ideal age for any form of extra-articular arthrodesis of the tuberculous hip is between eleven and twenty years. Before eleven years satisfactory results can be obtained but less frequently than in the ideal age group. It is also necessary to immobilise and splint these young patients for a much longer period than usual (Figs. 4 to 9).

RESULTS

Of the 147 operations, fifty-five were ilio-femoral grafts without osteotomy, and thirty-seven were ilio-femoral grafts followed by an osteotomy either within a few months to correct a pre-existing deformity or at a much later date to correct a deformity caused by failure of the grafting operation. Fifty-five patients were treated by an ischio-femoral graft combined with an osteotomy, as in the Brittain or Foley type of operation.

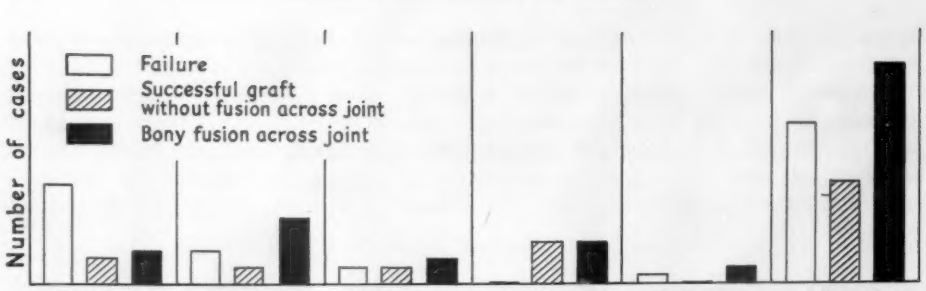

FIG. 4
Results of ilio-femoral graft without osteotomy.

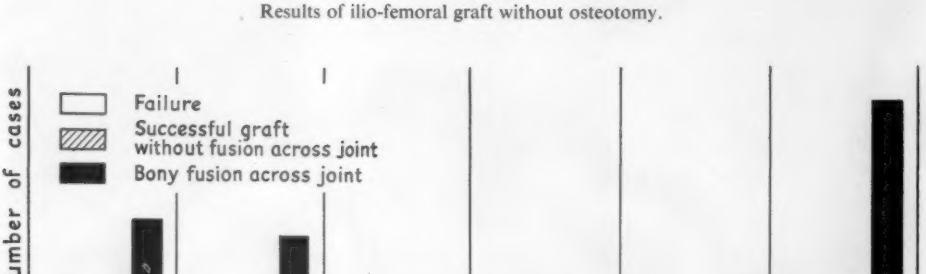

FIG. 5
Results of ilio-femoral graft with osteotomy.

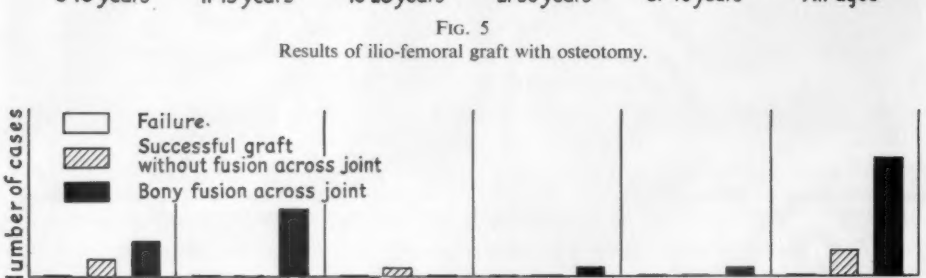

FIG. 6
Results of ilio-femoral graft with " early " osteotomy.

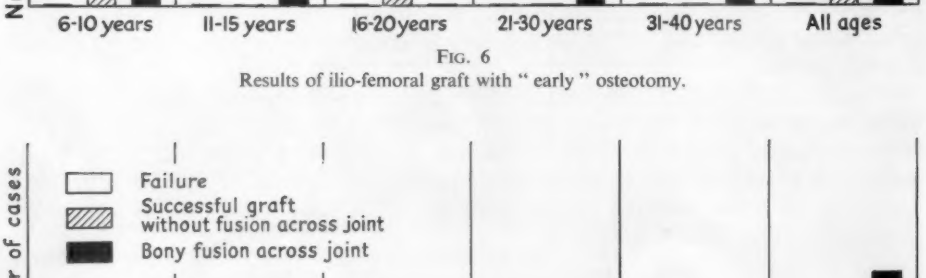

FIG. 7
Results of ilio-femoral graft with " late " osteotomy, related to age at time of osteotomy. In these cases the graft alone had failed.

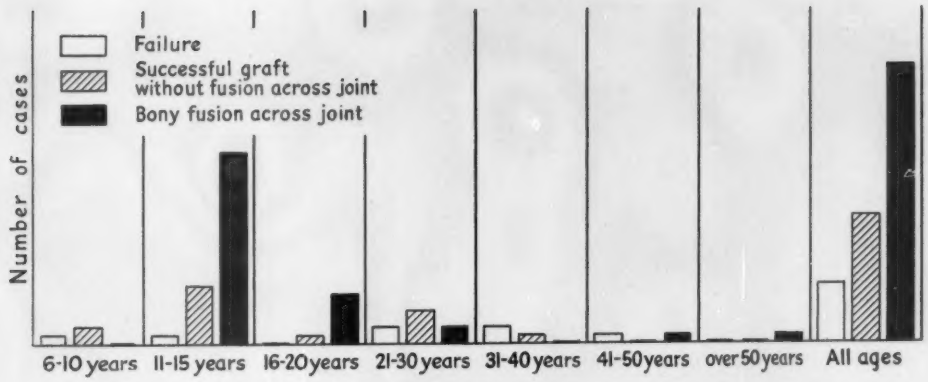

FIG. 8
Results of ischio-femoral graft.

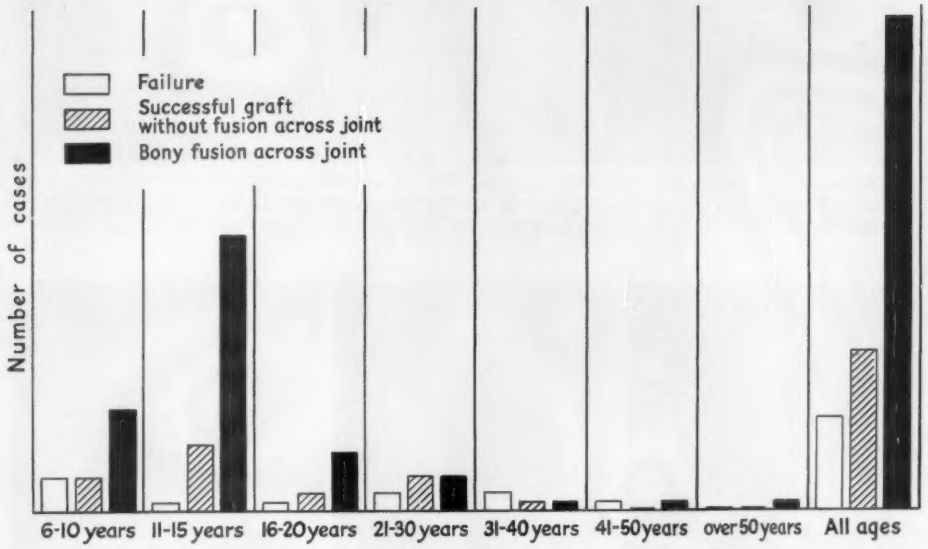

FIG. 9
Results in all cases of ilio-femoral and ischio-femoral arthrodesis combined with osteotomy.

Of these operations four—two ilio-femoral grafts without osteotomy and two ilio-femoral grafts with osteotomy—have been excluded owing to incomplete recent information. From these 147 operations there has been one death. This occurred in a child of ten subjected to combined ilio-femoral graft and osteotomy. This case has also been omitted from the results.

Thus 142 cases are available for review—fifty-three ilio-femoral grafts without osteotomy, thirty-four ilio-femoral grafts followed by an osteotomy of the upper end of the femur after a variable interval, and fifty-five ischio-femoral grafts of the Brittain or Foley type.

Of the fifty-three ilio-femoral grafts without osteotomy, nineteen were failures because the grafts failed to fuse at one or other end or became absorbed. In twelve cases a satisfactory extra-articular arthrodesis occurred, with an intact graft fused at both ends but without bony fusion across the joint. In twenty-two cases bony fusion occurred across the joint (Figs. 10 to 14).

FIG. 10

FIG. 11

Case 2. Figure 10—Quiescent tuberculosis of the left hip in a girl of eight. Figure 11—Nineteen months after ilio-femoral graft without osteotomy, showing absorption of the grafts opposite the joint line.

FIG. 12

FIG. 13

FIG. 14

Case 3. Figure 12—Quiescent tuberculosis of the right hip in a boy of twelve. Figure 13—Four months after ilio-femoral graft without osteotomy. Figure 14—At fifteen months the ilio-femoral grafts fused to the femur and ilium. Bony fusion also appears to be occurring across the joint.

FIG. 15

FIG. 16

FIG. 17

Case 4. Figure 15—Quiescent tuberculosis of left hip in a girl of twelve. Figure 16—Four months after ilio-femoral graft and immediately before subtrochanteric osteotomy. Figure 17—Twenty-seven months after the graft and twenty-three months after osteotomy, showing bony fusion across the joint. An example of ilio-femoral graft and " early " osteotomy.

FIG. 18

FIG. 19

FIG. 20

Case 5. Figure 18—Tuberculosis of the left hip in a girl of sixteen. The radiograph, taken ten years after ilio-femoral graft without osteotomy, shows a gap in the grafts opposite the joint. Figure 19—Four months after subtrochanteric osteotomy to correct flexion, adduction and lateral rotation deformity. Figure 20—Forty-two months after osteotomy, showing sound bony fusion across the joint and of the grafts.

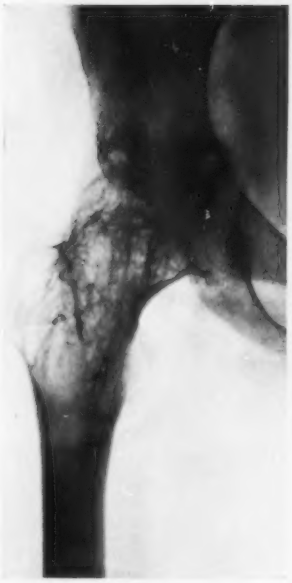

FIG. 21

FIG. 22

FIG. 23

Case 6. Figure 21—Quiescent tuberculosis of the right hip in a girl of eleven. Figure 22—Ten months after ischio-femoral graft by the posterior approach, showing soundly united graft and bony fusion across the joint. Figure 23—Five years after graft showing hypertrophy of the graft, sound bony fusion across the joint and atrophy of the great trochanter and lateral part of the femoral neck due to altered stresses.

Among the thirty-four ilio-femoral grafts followed by an osteotomy at an unspecified interval, there were four failures and four satisfactory extra-articular grafts without bony fusion across the joint. In the remaining twenty-six cases there was bony fusion across the joint (Figs. 15 to 20).

Of the fifty-five ischio-femoral grafts with osteotomy, seven were failures and fifteen were satisfactory grafts without promoting bony fusion across the joint. In thirty-three cases there was bony fusion across the joint (Figs. 21 to 23) (Table I).

TABLE I

RESULTS (ALL CASES)

Type of operation	Number of cases	Failure	Successful result		
			Fused graft only	Bony fusion across joint	Total
Ilio-femoral graft alone . .	53	19 (35·8%)	12 (22·6%)	22 (41·5%)	34 (64%)
Ilio-femoral graft with osteotomy	34	4 (11·75%)	4 (11·75%)	26 (76·5%)	30 (88%)
Ischio-femoral graft . . .	55	7 (12·7%)	15 (27·2%)	33 (60·0%)	48 (87%)

Thus thirty-four (64·1 per cent) ilio-femoral grafts without osteotomy, thirty (88·25 per cent) ilio-femoral grafts with osteotomy, and forty-eight (87·2 per cent) ischio-femoral grafts (Brittain or Foley type) resulted in successful extra-articular or intra-articular arthrodesis.

When the ilio-femoral grafts followed by osteotomy were subdivided into two groups, those with "early" osteotomy to correct pre-existing deformity, and those with "late" osteotomy to correct subsequent deformity, the results were as follows. Seventeen ilio-femoral

FIG. 24

FIG. 25

FIG. 26

FIG. 27

FIG. 28

Case 7—Tuberculosis of right hip, beginning at age three. Figure 24—Radiograph at age fourteen. Quiescent disease with a persistent irregular joint space. Figure 25—Four months after ilio-femoral graft using one piece of tibia. Figure 26—Twenty-eight months after operation showing absorption opposite the joint line, which is still visible. Figure 27—Four months after subtrochanteric osteotomy to correct adduction deformity. Union of the graft and commencing fusion across the joint. Figure 28—Sixteen months after osteotomy, showing hypertrophied ilio-femoral graft and bony fusion across the joint.

FIG. 29

FIG. 30

FIG. 31

FIG. 32

FIG. 33

Case 8—Tuberculosis of the right hip in a woman, beginning at age twenty-two. Figure 29—Radiograph at age twenty-five, showing dislocation of the joint. Figure 30—Six months later and three months after ilio-femoral graft, showing the graft along the upper surface of the femoral neck. Figure 31—Five months after subtrochanteric osteotomy carried out four months after ilio-femoral graft and "early" osteotomy showing satisfactory graft and bony fusion across the joint. Figure 32—Twenty-two months after ilio-femoral graft. Figure 33—Ten years later showing clearly the graft parallel to the upper surface of the femoral neck and sound bony fusion across the joint.

grafts followed by an " early " osteotomy resulted in three satisfactory grafts without bony fusion across the joint and fourteen cases with bony fusion across the joint; there were no failures. Seventeen ilio-femoral grafts followed by a " late " osteotomy resulted in four failures and one satisfactory graft without bony fusion across the joint; in twelve cases there was bony fusion across the joint (Table II).

TABLE II

RESULTS OF ILIO-FEMORAL GRAFT WITH OSTEOTOMY RELATED TO INTERVAL BETWEEN GRAFTING AND OSTEOTOMY

	Number of cases	Failure	Successful result		
			Fused graft only	Bony fusion across joint	Total
" Early " osteotomy . . .	17	0	3 (17·65%)	14 (82·35%)	17 (100%)
" Late " osteotomy . . .	17	4 (23·5%)	1 (5·9%)	12 (70·6%)	13 (76·5%)

Therefore seventeen (100 per cent) ilio-femoral grafts with " early " osteotomy and thirteen (76·5 per cent) ilio-femoral grafts with " late " osteotomy resulted in successful extra-articular or intra-articular arthrodeses.

The above figures suggest that femoral osteotomy is an important factor in successful extra-articular arthrodesis of a tuberculous hip. Further illustrative radiographs are shown in Figures 24 to 33.

COMMENT

In the ischio-femoral graft with osteotomy it is advisable to displace the shaft fragment medially to obtain support under the graft and to get a buttress of callus in this situation. In the ilio-femoral graft this medial displacement does not appear to be so important, whether the osteotomy is carried out early or late. If, however, there has been little destruction of the femoral neck medial displacement of the femoral shaft produces better alignment. It also reduces the shearing strain on the graft and the joint.

From these results it appears that, so long as an osteotomy of the upper end of the femur is performed, it does not matter, apart from the site of disease, whether an ilio-femoral or ischio-femoral graft is carried out. The ischio-femoral arthrodesis by the Brittain or Foley technique requires only one operation and there is no doubt, at least in children, that fusion can be surprisingly rapid. Against this, the operation—and particularly the Foley modification—is more severe than either an ilio-femoral graft or a subtrochanteric or intertrochanteric osteotomy.

SUMMARY

1. One hundred and forty-two cases of extra-articular arthrodesis of quiescent tuberculous hips with fibrous ankylosis have been reviewed.

2. The methods used were the ilio-femoral graft with and without osteotomy, and the ischio-femoral graft by the Brittain or Foley technique.

3. Success occurs more frequently when the grafting operation is combined with or followed by a femoral osteotomy.

4. It is suggested that this success is due largely to the increased immobilisation afforded by the osteotomy.

5. It appears that equally good results can be obtained with either an ilio-femoral or an ischio-femoral graft in these cases provided that an upper femoral osteotomy is also carried out, preferably at or soon after the grafting operation.

6. An upper femoral osteotomy will frequently convert an unsuccessful extra-articular hip graft into a successful one without further grafting.

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ARTHRODESIS OF THE OSTEOARTHRITIC HIP JOINT

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Degenerative osteoarthritis limited to one joint occurs more commonly in the hip than in any other joint because the femoral head is susceptible to both anatomical and physiological defects—to structural deformity of the bone and also to impairment of the blood supply. Congenital subluxation, developmental dysplasia, deficiency disease, endocrine disorder, fracture, epiphysial separation and dislocation of the joint, all may give rise to coxa plana, coxa vara, coxa valga, coxa magna or other incongruities in the shape of the articulating bones; while at the same time the vascular supply through blood vessels of the capsule and ligamentum teres is so precarious that avascular necrosis, osteochondritis and degeneration of articular cartilage may arise. It is not intended in this contribution to discuss the pathology of erosion of articular cartilage, the formation of osteophytes, thickening of the inferior aspect of the femoral neck, changes in bone texture, or the development of cysts in the femoral head and roof of the acetabulum, or to consider whether these changes arise from ischaemia or hyperaemia. That is quite another study. It is proposed only to remind ourselves that these degenerative changes occur very commonly in middle life at the ages of thirty to sixty years in one hip joint alone without general involvement of other joints or even of the opposite hip, and to consider the treatment of such degenerative changes by arthrodesis.

The first reason for choosing a simple clinical study of arthrodesis of the hip for the Thomas Fairbank Birthday Volume is that so much of the life of this great surgeon was devoted to clinical study of the problems of congenital dysplasia of the hip joint which without good early treatment causes degenerative osteoarthritis at the age of about thirty to thirty-five years.

The second reason for reviewing patients treated by arthrodesis of the hip joint after an interval of five to twenty-five years from the time of operation is that recent discussions and contributions have suggested a degree of despair and despondency that seemed surprising. We had thought that no orthopaedic operation was more gratifying to the patient, more satisfying to the surgeon, or more certain of restoring function; and yet it is being said that it is an operation of great technical difficulty, that it is unreliable, and that we cannot be sure of gaining sound fusion. It is said moreover that even if fusion is sound there will nearly always be pain in the back, often painful stiffness of the knee, and usually difficulty in sitting.

Consider for example these quotations from recent surgical literature: " bony union is difficult and doubtful "; " bone fusion is not easy to achieve "; " thirty-three arthrodeses were attempted and only thirteen were satisfactory "; " the first attempt to repair pseudarthrosis of the hip resulted in 50 per cent of failure "; " it is very difficult to ensure solid fusion "; " this arthrodesis shows a strong tendency to fibrous union "; " the pseudarthrosis rate for the entire group was 23 per cent "; " the mortality rate was 3·4 per cent and the rate of infection was 6 per cent "; " combining delayed union and failure gives 43 per cent of cases not proceeding entirely as the surgeon would wish "; " one failure for every two fusions is not a good recommendation for any operation."

As if that did not sound bad enough by reason of serious failure in the primary purpose of the operation we must go on to quote: " all of us are disturbed by the difficult problem of the stiff knee following many months of immobilisation "; " prolonged immobilisation in plaster-of-Paris causes stiffness of the knee "; " apart from failure of osseous union the main disadvantage is painful stiffness of the knee "; " we only used a short spica because we feared knee stiffness "; " there is danger of permanent stiffness of the knee and prolonged pain in the knee."

Then, lest we were not already frightened, we read: " when the hip is fused, excessive strain is thrown on the lumbar spine "; " patients complained of back pain when lying in bed unless a pillow was placed under the knee on the involved side "; " back pain was present in all age groups following arthrodesis " (Stinchfield and Cavallaro 1950, Taylor 1950, Howard 1950, Osmond-Clarke 1950, Smith-Petersen 1950, Judet 1950, Law 1952, Charnley 1953, Apley and Denham 1955, and others).

Many of these observations have no doubt been quoted from one paper to another on the basis of rumour alone without personal confirmation. In fact, the most acid critics of arthrodesis of the hip joint are those who never do it. Nevertheless the criticisms have been repeated with such frequency that one of us (W. C. R.) undertook a late follow-up of every arthrodesis of the hip joint done by the other (R. W.-J.). This independent study confirmed former impressions. It proved that fusion of the osteoarthritic hip can be relied upon with near certainty. *Far from there being one failure in every two fusions there was sound bone fusion confirmed radiographically in* 94 *per cent, a mortality rate of nil, and a recovery of painless knee movement to the right angle or far beyond in* 91 *per cent of patients.*

Analysis of the results of arthrodesis of the hip joint—At first it may seem surprising that one surgeon can report successful bone fusion in 94 per cent of patients whereas another refers to failure in 23 per cent (Stinchfield 1950), one to failure in 43 per cent (Charnley 1953), and another to failure of one in every two fusions (Apley 1955)—but these observations are misleading. The comments of Charnley, Apley and others misrepresent the statistical study of Stinchfield and Cavallaro (1950) which was a conglomerate of many different types of operation performed by many different surgeons for many different disorders of the hip. They reported on 117 patients treated by what was called " arthrodesis of the joint " by colleagues, associates and house-officers, including such varied procedures as Hibbs' extra-articular fusion with trochanteric graft, attempted fusion with an iliac graft but no internal fixation, intra-articular fusion with or without straight nails, ischio-femoral grafting of the Brittain type, nail fixation alone without exposing the joint, and " an insufficient number of other types of arthrodeses to be of statistical value." There were failures in from one-quarter to one-half of the cases. But many of these operations were archaic and of no more than historic interest; many were of a type that are now known to be useless; some were of the standard known to be essential. Why then should such a mixed review be quoted as the results of arthrodesis of the hip as we understand it today?

The series of patients reviewed in this contribution had arthrodesis of the hip joint by one surgeon who did the operation himself and supervised the after-treatment himself. It seemed better to exclude the many hundreds of other arthrodeses of the hip done at the London Hospital, the Robert Jones and Agnes Hunt Orthopaedic Hospital, the Liverpool Royal Infirmary, and the Royal Air Force Hospitals by colleagues or associates, with no more than a relatively remote control. This is an attempt to assess the results that are possible from one standard type of arthrodesis in the hands of one surgeon who himself is responsible from the beginning to the end.

Unfortunately the records of more than sixty patients were lost in the fire that destroyed the Robert Jones and Agnes Hunt Orthopaedic Hospital in 1948. We have also failed to trace a few others. Thus there remain 141 patients treated by arthrodesis of the hip joint who have been followed up at regular intervals, with a review of the result as confirmed five years and up to twenty-five years from the date of operation, the average period being eleven years. More recent cases have been excluded because a follow-up of at least five years is essential. We need only look at the results of nail fixation alone to realise that many patients gain relief for two or three years but with unfortunate failure later on. Indeed one patient treated by nail fixation was comfortable for *ten* years and only then had serious trouble. We should also look at the results of arthroplasty by prosthetic replacement of the upper end of the femur which were reported upon enthusiastically after two or three years but caused

grave despondency thereafter. No operation such as the Brittain V-arthrodesis with nail and graft, McMurray osteotomy, Girdlestone excision of head and neck of the femur, excision of upper end of the femur with Batchelor osteotomy, central displacement by the Charnley technique, or prosthetic replacement of the femoral head as advised by Judet, should be considered as proved in success unless the follow-up of results is from at least five to twenty or more years.

In reaffirming that sound bone fusion was proved in 94 per cent of cases by clinical tests and confirmed by radiographic examination it must at once be said that this relates only to patients with osteoarthritis treated by the standard operation of denuding and refreshening the joint surfaces together with the internal fixation of a nail, perhaps also of a graft, with the external support of a double plaster spica of full length for several months and certainly not less than four months.* Of these 120 patients there was need in four for a second operation after failure of the first. Similarly after open reduction for old traumatic dislocation with attempted arthrodesis, one of the two successful fusions was gained only

TABLE I

RESULTS OF SIMPLE FIXATION OF THE HIP BY NAIL, INTRA-ARTICULAR ARTHRODESIS OF THE HIP, AND OPERATIVE REPLACEMENT OF UNREDUCED DISLOCATION OF THE HIP (141 patients)

Fixation of the osteoarthritic hip by nail only (14 *patients*)		
Sound bony ankylosis	1	
Fibrous fixation—relief for ten years	1	
Fibrous fixation—temporary relief	11	
Died	1	
Intra-articular arthrodesis of the osteoarthritic hip usually with both nail and iliac graft (120 *patients*)		
Sound bony ankylosis	113	
Unsound union—failed	7	
Died	nil	
Degenerative osteoarthritis	102	Failed 7
Congenital dislocation	11	Failed nil
Ununited McMurray osteotomy	3	Failed nil
Failed cup arthroplasty	4	Failed nil
Open reduction of old traumatic dislocation of the hip with attempted arthrodesis (7 patients)		
Sound bone ankylosis	2	
Unsound—failed	3	
Died	2	

after a second operation, the first having failed. Unlike the view taken by Stinchfield in his reports we think that these should be classified as successful results and not as failures. It may be added that all these five patients with unsound fusion from the first operation gained sound union of the arthrodesis from a second operation after the grafting of cancellous iliac bone; there was certainly not failure in 50 per cent, as has been reported in other articles.

Fixation by nail alone—Two other small groups of cases must be considered separately. First, there are fourteen elderly patients who were treated many years ago by the insertion of a nail alone, the joint never being exposed. This operation, which has long since been abandoned, was not expected to fuse the joint. If sound bone ankylosis is to be considered the standard of success these cases represent a failure of nearly 100 per cent. But obviously they should not be included in a statistical review of arthrodesis of the hip, as Stinchfield did, because the operation was never expected to be an arthrodesis.

Operative reduction of old traumatic dislocation of the hip—The second group to be considered separately are patients with traumatic dislocation of the hip joint, unreduced after many

* This review includes only patients with osteoarthritis of the hip joint of congenital, developmental, traumatic or paralytic origin, and none with tuberculous or acute suppurative arthritis.

X1(4)

months or years, where the essential measure was late operative replacement. Any attempt to secure fusion of the joint at the end of such an operation must surely be regarded as a supplementary procedure. In fact, the results in this small group of seven patients were so bad as to dissuade most surgeons from attempting operative reduction of old traumatic dislocations. Of seven patients so treated, two died, three developed pseudarthrosis, and only two gained sound fusion, one from a second operation after failure of the first. If these are to be included in the results of arthrodesis of the hip they show success in only 28 per cent, and failure in 72 per cent. Such failures are not however representative of the results of arthrodesis of the hip for degenerative osteoarthritis, congenital subluxation, epiphysial coxa vara, old Legg-Perthes' disease or paralytic subluxation.

FIXATION OF THE OSTEOARTHRITIC HIP BY NAIL ALONE

Nearly twenty years ago there was recorded an attempt made in elderly patients with osteoarthritis to fix the hip by the insertion of a nail from below the trochanter without exposing the joint (Watson-Jones 1938). The idea was that patients aged from sixty to eighty years who might not withstand the risks of an open intra-articular arthrodesis could perhaps be relieved of pain by the simple fixation of a nail, supplemented at first by a short plaster spica. We are sorry that this operation was ever recorded. It should have been obvious from the beginning that nail fixation alone could never sustain firm and sound fixation. Sooner or later, in consequence of the tremendous strains and forces that are concentrated at this level, there was bound to be resorption of bone with gradual loosening of the nail. Of the fourteen patients so treated, only one who had arthritis of a rheumatoid type gained sound ankylosis. One other, whose joint was already almost completely stiff at the time of the nailing, had relief for as long as ten years (Fig. 1) but even he had recurrence of severe pain after that—which shows how useless it is to assess the end-results of such operations after a follow-up period of only two or three years. In eleven there was temporary relief of pain lasting for a year or two but not longer. One died from fat embolism.

The ages of these patients ranged from sixty to seventy-six years. On late review it is obvious that the benefit from nailing alone was short-lived and that the procedure was hardly worth while. We have long since abandoned the operation. Unfortunately however it is more difficult to withdraw a recommendation than to propose it, and in various parts of the world bigger and better nails, stronger bolts, and more powerful screws and plates are still being used in an endeavour to fix the hip by an extra-articular method relying upon metal alone. They will all fail. Many patients of this age are of course too feeble and fragile to put up with the rigours of intra-articular arthrodesis with prolonged immobilisation in plaster. It is very possible that in this age group the Charnley operation of central displacement of the femoral head will find its right application. Certainly there is no merit in trying to fix the hip joint by a nail without intra-articular fusion, no matter how powerful the nail may be nor how strong the screws and bolts attached to it may be.

In acknowledging that a nail alone cannot permanently fix the hip we must also recognise that even after open exposure of the joint with complete denuding of articular cartilage, internal fixation, and the support of a plaster spica for several months, the nail itself cannot protect unsound consolidation. If the joint is not already soundly fused the nail will gradually loosen from resorption of bone wherever there are compression forces. The degree of such compression, and strength of the rotational forces, may be judged from the patient whose radiographs are shown in Figure 2. This girl had residual paralysis from poliomyelitis. A difficult decision had to be taken as to whether she might be improved by arthrodesis of the hip. It was decided, perhaps foolishly, to try the temporary insertion of a nail by which to assess function after stiffening the hip with the intention of withdrawing the nail without too much damage if the patient was made worse, but going on to formal arthrodesis if she was made better. This case is quoted only to show how great are the stresses at a stiffened

hip joint. Within a week or two of the trial operation and attempted weight bearing the joint was distracted and a heavy steel nail was twisted through 45 degrees. These are the forces with which we must contend.*

FIG. 1

This is the only patient who gained more than temporary relief from simple nail fixation of a painful osteoarthritic hip, his joint having already been almost completely stiff. He was happy for ten years—but let it be noted that as long as ten years was needed to determine the real end-result. We must not judge the results of operations on osteoarthritic hip joints from two or three year follow-up studies. Five to twenty or more years are needed to judge the end-result.

Look now at Figure 3. Here is a patient whose hip joint had been arthrodesed by one of our colleagues in Liverpool with full exposure of the joint, denuding the articular cartilage, and internal fixation by a three-flanged nail with protection in a plaster spica for three months. It was then thought that although the fusion was not quite sound it would be enough to rely upon the well placed nail for consolidation to be completed. Far from this being so, the unsound union became less sound, the fibrous fixation became less firm, mobility increased, and as months went by the pressure-resorption of bone permitted a range of movement of almost 90 degrees, the nail lying loose both in the pelvis and in the femoral neck (Fig. 3).

INTRA-ARTICULAR ARTHRODESIS OF THE OSTEOARTHRITIC HIP WITH NAILING AND GRAFTING

Before considering the results of intra-articular arthrodesis of the hip joint as reported in this series we must first discuss the indications that were insisted upon before the operation

* With considerable difficulty the twisted nail was removed and the joint was arthrodesed. Since then she has done well. The Trendelenburg limp and lurch was entirely controlled and her function was greatly improved. We regret only that we bothered her with the " trial nail fixation " which gave a lot of unnecessary pain and discomfort.

FIG. 2

This patient had residual paralysis from poliomyelitis and the question arose as to whether fusion of the hip might help in improving her function. A nail was inserted experimentally. Within a week or so of walking the radiograph, and the photograph of the extracted nail, show how very great is the leverage at a hip joint which is stiffened. A strong steel nail has been twisted through 45 degrees. These are the forces with which we must contend in attempting arthrodesis of the hip.

FIG. 3

Even after intra-articular arthrodesis and support in plaster for three or more months a nail does not suffice to protect unsound fusion, as proved in this case treated by one of our colleagues in Liverpool where a tremendous range of mobility developed. It was thought that early union might proceed to final consolidation by the protection of the nail—but see what happened. A nail can never be more than an internal suture.

was done. It should be emphasised that no patient was ever persuaded against his will. Not until it is quite certain that the degree of disability, and the severity of pain and stiffness, really warrants it should the hip be arthrodesed. This series of 141 patients is from a total of many hundreds of others with osteoarthritis of the hip joint who were advised that although fusion might be needed one day they were for the moment much too good for it. Many had pain and limitation of activity but still with about 90 degrees of movement and an ability to walk several miles, perhaps with a stick. They needed no more than reassurance. Many were worried and anxious because having been told that they had arthritis they feared crippling, invalidism and a bath-chair life. For them the essential treatment was reassurance that the " arthritis " was no more than a simple age change just like a hinge in a door which had begun to creak and squeak, and that it had nothing whatever to do with rheumatoid arthritis or any crippling disorder. Palliative measures such as massage, electrical heat, injection of lactic acid, the use of cortisone preparations, or irradiation of the joint may satisfy them that something is being done; but we think on the whole that it is better not to mislead patients by suggesting these measures which can never give permanent relief and are only temporary palliatives. If they want pain to be soothed by massage they can just as well rub the joint themselves without travelling long distances to departments of physical medicine. If they find that discomfort is relieved by heat they can just as well do it for themselves by a hot water bottle as by short-wave diathermy. The benefit of injection of lactic acid, hydrocortone, or other preparations have never been proved. The essential measure is to reassure the patient, advise the simple use of salicylates such as Asprin, Disprin, or Veganin and to be told that if, as years go by, there is increasing pain and stiffness the symptoms can always be controlled by a stabilising operation.

When pain and stiffness of the hip joint becomes so disabling that there is difficulty in walking more than a few hundred yards, or at the most a mile, arthrodesis should be advised. An important practical point at once arises. If it is put to the patient that the operation will be a stiffening procedure it is more than likely that it will be refused. He says to himself " the joint is very stiff now; what will it be like when he has finished with it? " It should not be put as a stiffening operation but rather that it is a stabilising operation. It should be explained that after successful fusion there will still be a range of movement that looks and feels like hip movement up to 30 or 40 degrees in every direction. The range of apparent movement, which of course is pelvic movement, should be demonstrated, it then being explained that with such mobility there will be every facility in walking without an appreciable limp, sitting comfortably, climbing, running, jumping, and indeed pursuing every activity. Many patients still have apprehension in wondering how a stiffening or stabilising operation can permit such apparent mobility and energetic activity. They should meet other patients who have had an arthrodesis of the hip. Of all operations that may have to be suggested none is more difficult for the patient to understand than this. The fact of the matter is that those who have got over initial doubts and had their hip joints arthrodesed are the first to suggest that they would like to welcome others and show what comfort, relief of pain, restoration of usefulness and energetic activity is possible.

Indications for intra-articular arthrodesis of the hip joint—Intra-articular arthrodesis is not indicated unless there is: 1) considerable pain in the hip, thigh and knee; 2) movement restricted to about half of the normal range or less; 3) restriction of walking to about one mile or as little as a few hundred yards; 4) unilateral arthritis, the opposite hip having a normal range of movement including almost full internal rotation, limitation of which is of course the first sign of degenerative osteoarthritis; 5) free mobility of the knee joint on the side of the affected hip. The success of arthrodesis of one hip joint depends upon free mobility of the other. If there is osteoarthritis of both hips some type of arthroplasty, osteotomy, or other mobilising procedure is necessary; but if the opposite hip is normal there need be no hesitation in arthrodesing the affected one no matter whether the patient is aged forty,

fifty or even more than sixty years. Occasionally it may be wise to arthrodese one hip joint even when there are early signs of arthritic change in the opposite hip in order that the joint that is more advanced in its degenerative change may become " the good joint " which will take all strains and thus protect the other.

Contra-indication of arthrodesis when the knee joint of the same limb is already stiff—We should be very cautious in advising arthrodesis of the hip joint when the knee on the same side is already stiff. Ability to walk well and sit comfortably after arthrodesis of the hip depends on free mobility of the knee joint of the same limb.

No contra-indication of arthrodesis of the hip by fear of low back pain—Pain in the low back, even when accompanied by radiographic evidence of spondylosis or degenerative arthritis of the intervertebral joints, is not necessarily a contra-indication to arthrodesis of the hip. It is very often said that fusion of the hip gives rise to pain in the low back, but this is quite untrue. If an arthritic hip joint is fused in the neutral position so that external rotation and adduction deformity are corrected low back pain is relieved. Quite unlike the observations so commonly made and reported in the literature, arthrodesis of the hip joint does not cause pain in the back. Low back pain arises from strain of the intervertebral joints from unsound fixation of a hip in a deformed position. When fixation of the hip is sound, and the deformities are corrected, such low back pain as there had been is made better and not worse.

Age of the patient in determining whether to arthrodese the hip—Finally we must consider age. By far the greatest number now reported were aged from twenty to sixty years (ninety-nine patients—just over 81 per cent). It is obvious that there may be caution in advising such an operation for patients who are aged more than sixty years because many of them do not want to be able to walk ten or twenty miles, go mountaineering, climb ladders, or run and jump. They may well prefer a palliative procedure such as arthroplasty or osteotomy which gives them all they need, with ability to sit elegantly and comfortably. Nevertheless, the decision as to whether or not a hip joint should be arthrodesed must be based on the physiological age of the patient and not on the dictates of a calendar. Many men and women of sixty or more years are still energetic and athletic, and they *do* want to be able to walk unlimited distances, never considering whether to take a walking stick, always forgetting that they have any restriction of activity. In this series of 141 patients no less than twenty-eight were aged from sixty to eighty years (23 per cent of the series). We well remember the patient who sustained a fracture of the neck of the femur at the age of ninety-two who had never forgiven a doctor who thirty years earlier, when she was aged sixty-two, had said " you are too old for an operation."

Only seven patients were aged less than twenty years (5·8 per cent). It so happens that none were in the first decade where suppurative arthritis of infancy may best be dealt with by arthrodesis as recorded by Bryson (1948). In this particular series all seven patients were in adolescent years, their hip disability arising from congenital subluxation, paralysis of muscles, Legg-Perthes' disease, or deformity of the femoral head from fracture of the neck. We feel strongly that in these patients the benefits of arthrodesis should not be withheld just because they are young. Figure 15 shows the radiograph of the hip of a girl aged fifteen who two years before had sustained a fracture of the femoral neck. As so often happens when the neck of the femur is fractured in adolescent years there was avascular necrosis of the femoral head. It had been proposed to remove the nail, replace it with a graft from the fibula, and correct the apparent shortening of more than two inches by a femoral osteotomy. In giving this advice the surgeon had said that arthrodesis would surely be needed when the patient reached the age of about thirty years but proposed a palliative operation meanwhile. What benefit is conferred by changing a total cripple to half a cripple in the young years of life that matter most? Why not in such circumstances go at once to the arthrodesis of the joint that is inevitable?

TECHNIQUE OF INTRA-ARTICULAR ARTHRODESIS OF THE HIP JOINT

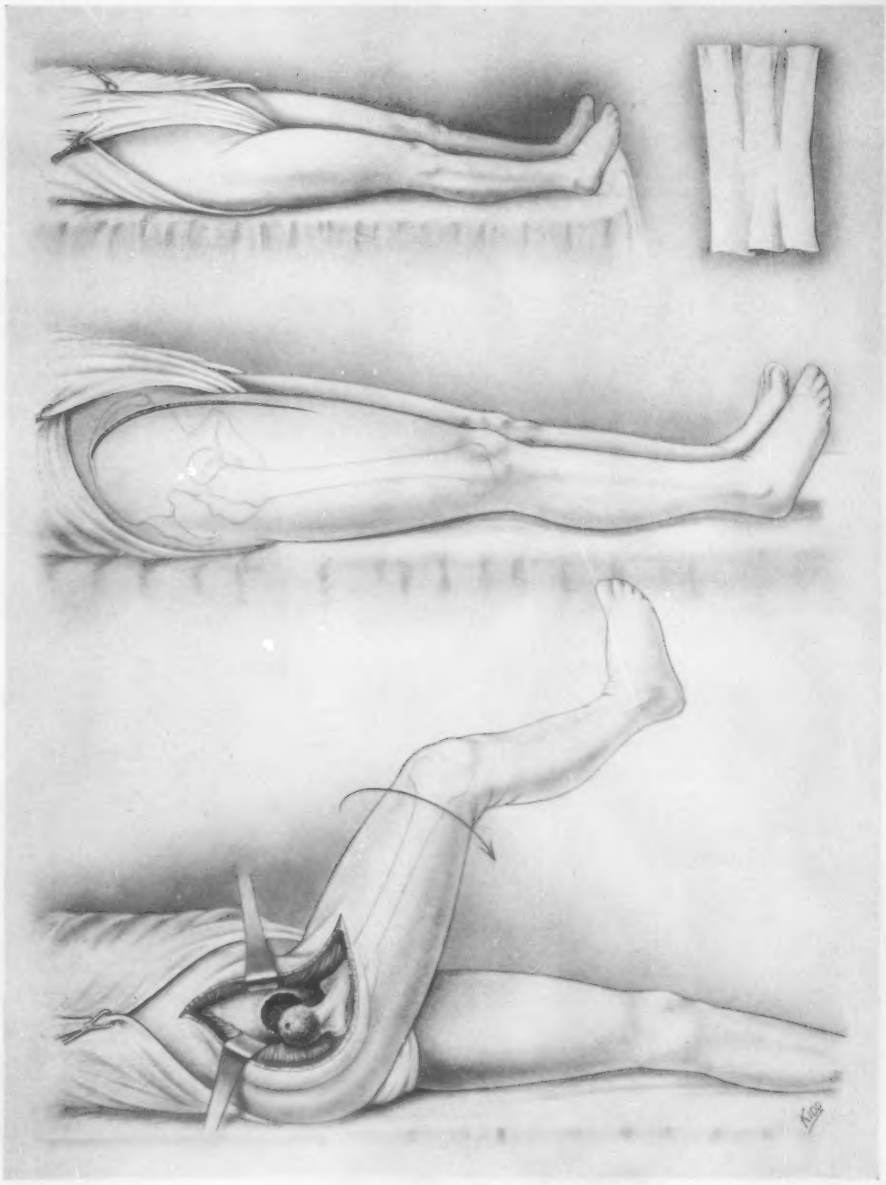
It must be admitted that arthrodesis of the hip joint is by no means an easy operation. There are technical difficulties; but with reasonable skill by the surgeon, and the care of a good anaesthetist who may sometimes want to use a saline drip and very occasionally transfusion of blood, the risks of this operation are less than those of walking across Piccadilly Circus. At the end of it there should be no more than slight staining of the side towels, and certainly no more loss of blood than about half a pint. Preliminary infiltration of an adrenalin-hyalase mixture is wise, as well as the use of diathermy for quick control of small bleeding vessels. The only important group of blood vessels that need ligation are the lateral circumflex arteries.

Exposure of the joint and removing articular cartilage—The Smith-Petersen incision is best. After separating the gluteal muscles from the dorsum of the ilium and inserting a pack, the front of the capsule is excised. The limb must then be rotated outwards through at least 90 degrees in order that the whole of the acetabulum may be seen quite easily. Every fragment of cartilage is removed from the acetabulum with the aid of a long gouge, preferably about twelve inches in length because in fat patients and those with well developed muscles a shorter gouge can almost be lost in the depths of the wound. The articular cartilage of the femoral head is then removed. A small incision is made over the lateral aspect of the femur, just below the greater trochanter, through which to insert a guide wire traversing the neck of the femur until its point is seen in the upper part of the femoral head. The length of guide wire inside the bone can be determined by measuring the length of wire still outside the bone and subtracting it from the known total length of the wire. A nail of the correct length is chosen—about one inch longer than the penetrated part of the guide wire so that when it is driven in a full inch will engage in the pelvis (Figs. 4 to 11). But first, one of the most important parts of the operation arises.

Position of the fused hip—Successful function will be achieved only if the hip is fused in a position of neutral rotation with no more abduction than is needed to correct true shortening. The total apparent lengths of the limbs should be exactly equal. In earlier years some surgeons advised that the hip joint should be fused in an externally rotated and abducted position—but this of course is nonsense. Many imperfect results that have been reported have arisen from this out-dated view. The limb must be in exactly neutral rotation with the patella pointing to the ceiling. There must be no more abduction than is needed to correct true shortening, which is seldom more than about 10–15 degrees. There must be no deliberate flexion of the limb. As the patient lies on the table there is enough lordosis to be sure that the final position of consolidation will amount to only 30 degrees of fixed flexion which is quite enough for comfortable sitting and is correct for comfortable standing.

Pull-push clamps—In order to do this it is helpful to use the clamps illustrated in Figures 12 and 13. They are sterilised and screwed over the sterile towels at the end of the table, the feet of the patient being fixed to them with sterile bandages. The total apparent limb length is then corrected by pushing on the normal limb and pulling on the affected limb until the malleoli are exactly level. Care must be taken to see that the apparent equality of leg length has really been achieved by sufficient slight tilting of the pelvis and not by distraction of the femoral head from the acetabulum on the side of the hip that has been exposed. The guide wire is then driven into the pelvis and the nail is punched over it. The final stage of the fusion is to impact fragments of cancellous bone cut from the ilium.

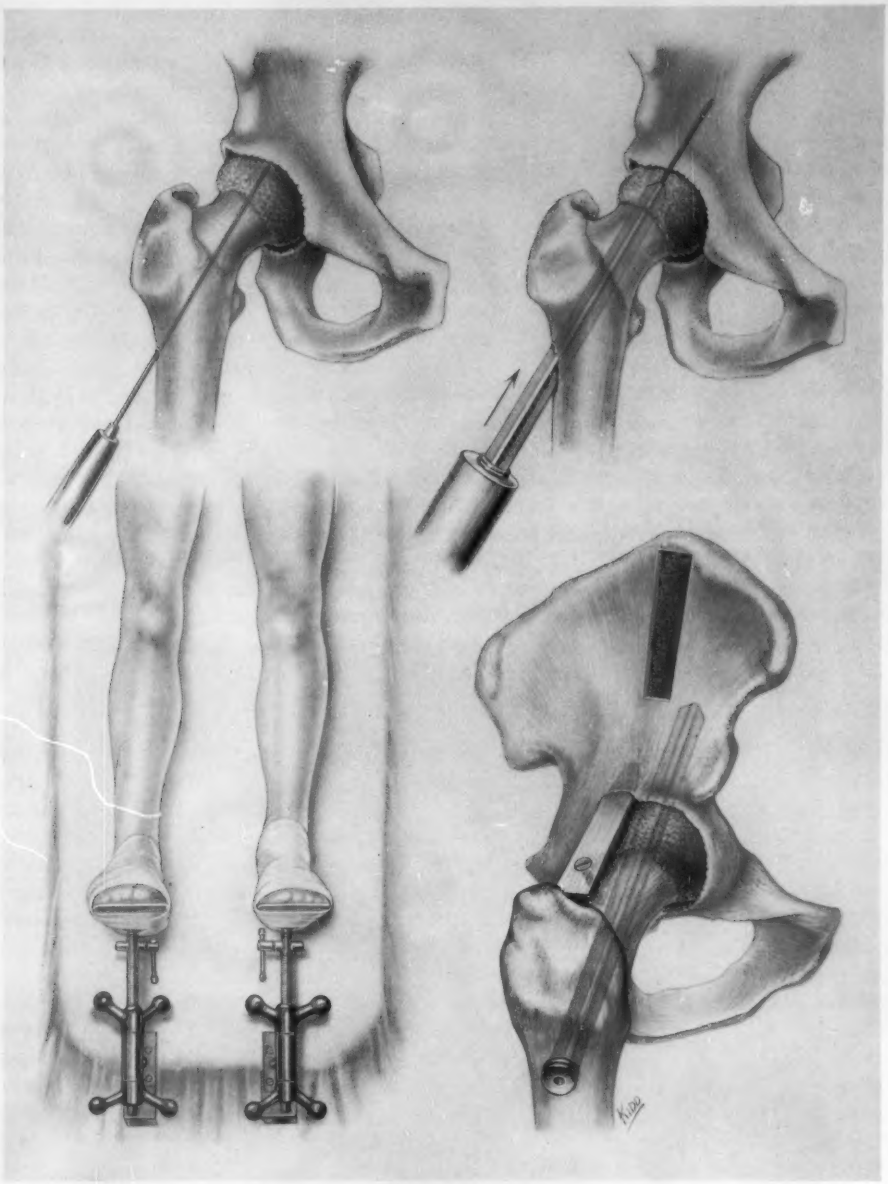
Insertion of cancellous iliac bone—Any surgeon who does this type of fusion of the hip joint often enough knows that even after completely denuding the articular surfaces of the acetabulum and femoral head it is quite difficult to find enough space into which cancellous chip grafts of bone can be punched. It is wrong to say that " the mechanical defect of the Watson-Jones method is the lack of congruity between the enlarged acetabulum and the reduced size of the head; even when packed with bone chips the head touches the acetabulum



FIGS. 4–7

Technique of Arthrodesis of the Hip Joint

Special towelling is needed so that both lower limbs, covered with sterile stockinette, are exposed from the feet to the pelvis in order that there may be accurate adjustment of length. A Smith-Petersen incision is made. An essential point of the exposure is full external rotation of the limb through at least 90 degrees so that the whole of the acetabulum is seen, as well as the femoral head. This considerable rotation of the limb must of course be done with sufficient caution to prevent fracture of the neck or upper shaft of the femur.



FIGS. 8–11

Technique of Arthrodesis of the Hip Joint

After the cartilage of the acetabulum and femoral head has been denuded a guide wire is driven from below the trochanter until its point emerges in the upper part of the femoral head. The position of the limbs is then adjusted by special clamps fixed to the lower end of the table so that the total apparent lengths of the limbs are equal, true shortening being corrected by slight tilting of the pelvis with abduction of the affected limb. A nail is then driven over the guide wire into the pelvis. Finally a graft cut from the ilium is slotted into the roof of the acetabulum and screwed to the neck of the femur.

FIG. 12

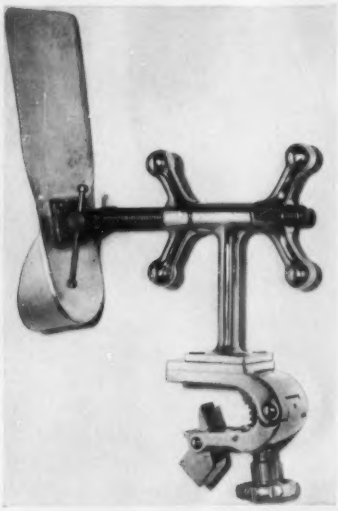


FIG. 13

This demonstrates the clamps which are sterilised and fixed over the end of the table and bandaged to both feet so that by pull and push the total apparent lengths of the lower limbs is exactly equal, the malleoli being level.

FIG. 14

If the nailing is done at a separate second-stage operation it is better to use a Watson-Jones traction table which makes it easy to get the malleoli exactly level, correcting true shortening by sufficient abduction of the hip and facilitating the immediate application of a plaster spica.

over a small area of contact." It is still worse to say that " the well known analogy of the orange and the cup cannot be better—before operation the head is like an orange inside a cup, whereas after the operation it is like a peeled orange lying on a saucer " (Charnley 1953). This is a complete travesty of the facts of the operation.

After the nail has been driven into the pelvis the femur should be impacted closely into the acetabulum by hammering over an impactor punch in the trochanteric region. Then it is usually wise to cut a whole thickness graft from the dorsum of the ilium, slightly wedged in shape, the narrower end being driven into a slot cut in the bone just above the acetabulum, and the other end fixed to the femoral neck with one screw (Figs. 20 and 21). At the end of the operation the internal fixation provided by the nail and bone graft should be absolutely sound and firm so that there is no rock at all between femur and pelvis when the limb is moved. The nail itself often provides such a degree of sound fixation that an onlaid iliac graft is not essential (Figs. 18 and 19). If, however, after driving the nail and impacting the femoral head into the acetabulum, there is still slight mobility a graft should be used (Figs. 23 and 24).

It is because of the need for complete internal fixation that during the last ten years we have used a one-stage and not a two-stage operation. Before that, because of the fear of surgical shock after arthrodesis, we used a two-stage operation in which all articular cartilage was first denuded and a plaster spica applied—a procedure that takes no more than about twenty minutes operating time and twenty minutes for the plaster—with the insertion of a nail at a second-stage operation two or three weeks later. This, however, gives no opportunity for noting the degree of success of the internal fixation. Now that anaesthesia has become so good, and the risk of surgical shock is so low, a one-stage operation is done. The denuding of articular cartilage, correction of total apparent length of the femur, insertion of a nail, and onlaying of a bone graft, are all done at the same time, the operating time being in the region of sixty minutes.

AFTER-TREATMENT OF ARTHRODESIS OF THE HIP JOINT

Duration of immobilisation in plaster—The one essential point that matters in after-treatment is that the limb must be immobilised in a double plaster spica, as far as the toes on the affected side and to just above the knee on the opposite side, for not less than four months. Every attempt that has been made to lessen this annoying and irritating period to three months, twelve weeks, ten weeks, or even less, has caused failure of sound fusion in increasing numbers of patients in direct proportion to the time that the duration of immobilisation has been reduced. It takes at least four months for osteoid and callus to consolidate. Sufficiently strong bone cannot be established in less time than that after fixation of a hip joint where there are such stresses and strains. Unless the patient knows from the beginning that four months of immobilisation in plaster will be needed, and possibly a little longer, it is better not to attempt arthrodesis of the hip joint. Nearly always, although the price is heavy, the patient accepts this. After all, what does four months of relative immobility in bed matter in comparison with the twenty, thirty or more years of happiness that will be enjoyed thereafter? As a rule the plaster spica applied at the time of operation is changed for a more closely fitting cast after a few weeks. The patient can then go home or to a convalescent centre. After four months, clinical tests are made and radiographs are taken to confirm the soundness of fusion. If there is doubt, another plaster spica should be applied for one or two months longer. When it is sure that fusion is sound the knee joint is mobilised by simple active exercise and the patient is taught to walk without a limp, and finally is instructed in running, jumping, climbing or any recreation in which he is interested.

Mobilisation of the temporarily stiff knee joint—Although after four or more months of immobilisation in plaster the knee joint will be temporarily stiff, a free range of movement will be regained provided only that strenuous physiotherapy, passive force and manipulation

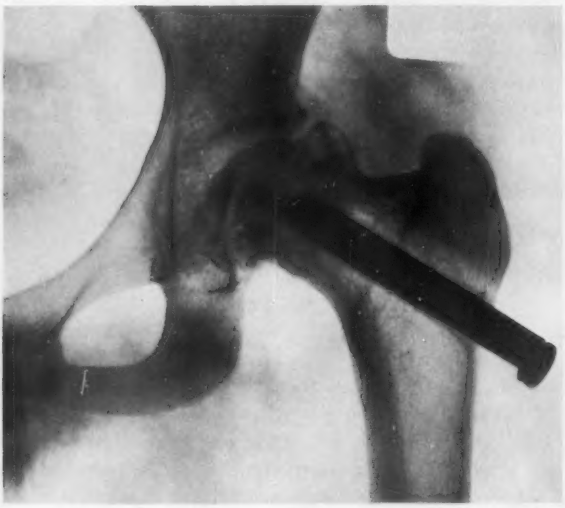


FIG. 15

This girl, aged fifteen years, sustained a fracture of the femoral neck. It was immobilised by a nail but as so often happens at this age she developed avascular necrosis of the femoral head with sequestration of about half of it. There is gross irregularity in the shape of the bone. It is obvious that degenerative arthritis will develop by the age of thirty-five years. Why not then go straight to the arthrodesis that will be needed rather than attempting a palliative operation just because she is aged no more than fifteen years?

FIG. 16

FIG. 17

Very occasionally, as in this case of congenital dislocation of the hip, it may be possible to secure such firm fixation of the femoral head in a hollowed area of the pelvis, together with an iliac graft, as to make the use of a nail unnecessary.

FIG. 18

FIG. 19

Arthrodesis of the hip joint in a patient with an old displaced upper femoral epiphysis. Internal fixation was secured by a three-flanged nail. This is the patient shown in Figure 31 who is rock climbing.

FIG. 20

FIG. 21

After denuding the joint surfaces and using the internal fixation of a nail an iliac graft is usually driven into the pelvis just above the acetabulum and secured to the femoral neck with one screw. These two cases illustrate the technique.

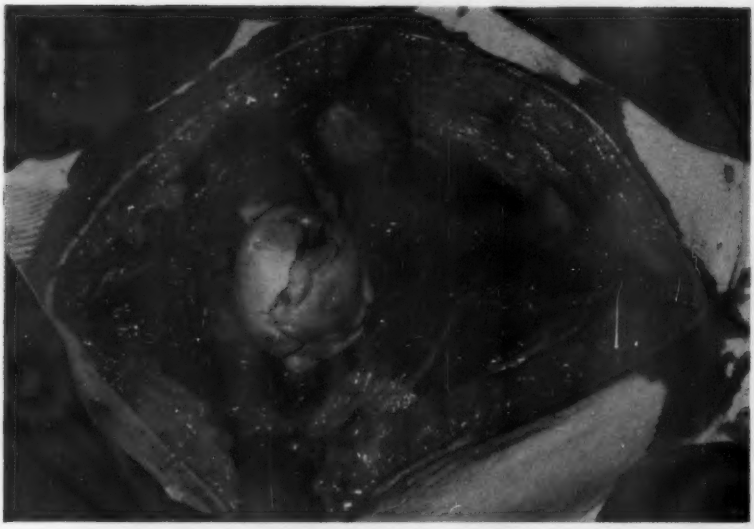


FIG. 22

After exposure of the joint through a Smith-Petersen incision the limb is externally rotated strongly, usually through 90 degrees, so that the acetabulum is easily seen. Note in this case of a young girl the extent of sequestration of two-thirds of the femoral head (see Fig. 15). This dead fragment could be lifted out with a pair of forceps.

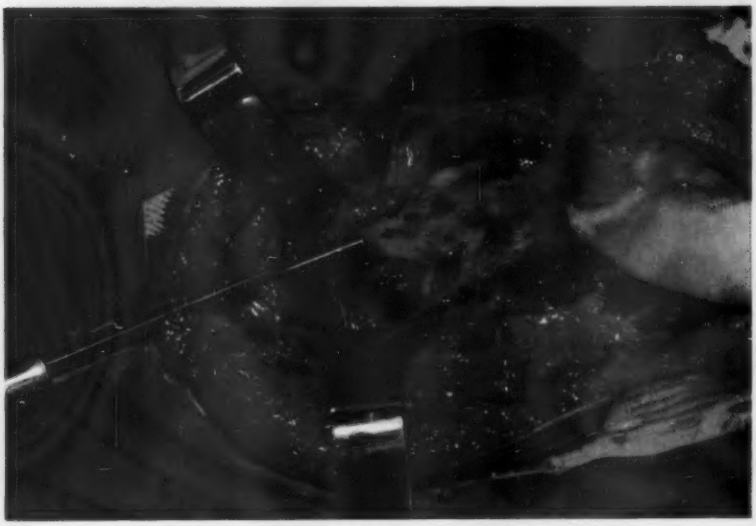


FIG. 23

After denuding all articular cartilage the femoral head is replaced in the acetabulum, a guide wire is driven in from below the trochanter and seen to emerge in the upper part of the femoral head. Then, after adjustment of the limb lengths, the wire is driven on into the pelvis. This illustration shows the nail being driven over the guide wire.

are avoided. The quoted observations from the literature as to the dangers of permanent and painful stiffness of the knee arise largely from surgeons who do not know how to treat a stiff knee. Like every other stiff joint, particularly for example the fingers, elbow and shoulder, so too a knee joint that is temporarily stiff will become permanently stiff if it is irritated and worried by repeated stretching, passive force and violence. But if the patient is encouraged, persuaded and cajoled into his own regular exercise for not more than five minutes at a time, but repeated every hour of the day, the joint will regain at least as much movement as there was before the operation.

Protection from oedema and restoration of the normal circulation—At first the limb that has been in plaster for many months should be supported with a crepe bandage or elastic stocking until the tone of muscles is restored, so that the venous return becomes normal and the tendency to oedema is controlled.

FIG. 24

Finally a thick iliac graft is punched into a slot in the bone just above the acetabulum and fixed by one screw to the femoral neck.

Summary of the operative treatment and after-treatment—Before analysing the late follow-up results of patients treated by arthrodesis of the hip joint we would emphasise that in this series there was: 1) complete exposure of the joint with denuding of articular cartilage from the acetabulum and femoral head; 2) internal fixation by means of a nail driven from below the trochanter into the pelvis, or an iliac graft, or usually both; 3) immobilisation in a double plaster spica for at least four months; 4) mobilisation of the knee joint by active exercise alone without passive stretching or manipulation.

RESULTS OF ARTHRODESIS OF THE HIP

Every adversity to the patient that arose within five years of operation has been noted, many not really being related to the arthrodesis itself. For example one had cerebral thrombosis with hemiplegia on the opposite side after twelve months, but still says that the operation was worth while because that limb is now the stronger one by which she can protect the more recently paralysed limb. Two others who died at five months and six months after operation have been excluded from the statistical review because the death of one was from acute appendicitis and the other from a self-administered overdose of barbiturates.

Complications of the operation—Two patients sustained subtrochanteric fracture of the femur, one after twelve months and one after two years. There seems no doubt that fusion of the hip joint, by reducing mobility at this level, predisposes to this fracture just below the level of the head of the nail. Two had drop-foot, noted at the time that the plaster was removed, which recovered fully within a few months. It seems probable that the paralysis arose from pressure of the plaster on the lateral popliteal nerve where it winds round the neck of the fibula. One other had a pulmonary infarct at the fourth week from which she recovered. In four there was thrombosis of the femoral vein, two with very troublesome and persistent swelling and oedema of the limb needing the permanent use of an elastic stocking. This complication is quite probably related to the insertion of retractors on the medial side of the joint in exposing the depths of the acetabulum; certainly there should be great care and gentleness in the introduction and holding of such retractors.

Mortality—Of the 141 patients in this review three died. One was an elderly lady who had nail fixation alone and died on the fourth day from fat embolism. The other two had unreduced traumatic dislocations of the hip joint, one dying from surgical shock within twenty-four hours and one from a mismatched blood transfusion. Thus of fourteen patients treated by nail fixation alone there was one death; of seven treated by operative reduction of old unreduced traumatic dislocation of the hip there were two deaths; and of 120 patients with osteoarthritis of the hip joint treated by intra-articular arthrodesis there were no deaths. The safety of any operation of this magnitude does of course rest just as much upon the anaesthetist as upon the surgeon. The surgeon must control bleeding, minimise tissue damage, do no more hammering than is essential, and not waste time. But whereas in former years many pints of blood were transfused we now rely upon the skill of anaesthetists; blood transfusion is hardly ever needed.

Wound infection—Of the 141 patients who were operated upon there was wound infection in five (3·5 per cent), this being based upon a stringent and rigid assessment which means that superficial infection or an infected haematoma was so recorded. In none of these five did infection reach the bone; in none was there interference with bone fusion; and in none did the infection give rise to more than temporary anxiety. Penicillin, terramycin, chloromycetin or other antibiotics were used only occasionally as an umbrella control. We did however always use non-touch technique.

Stiffness of the knee joint—The number of patients who regained less than right-angled flexion of the knee joint was twelve (8·5 per cent). Such limitation of knee movement is of course unfortunate after arthrodesis of the hip because it causes difficulty in sitting, and annoyance in putting on stockings or reaching shoe laces. Over 90 per cent regained comfortable and painless movement of the knee joint to the right angle or far beyond. In 40 per cent of patients of all age groups, even up to sixty-five or seventy years and after immobilisation in a full plaster spica for at least four months, there was normal movement so that the heel could touch the buttock. Figures 26 to 30 show a typical sample of the ranges of knee movement that may be expected. For the few patients whose knee joints remain stiff there are many simple devices to aid in dressing such as tapes attached to the top of stockings, elastic laces for shoes, and long-handled shoe-horns.

Ability to sit comfortably—Look at Figure 25. It is often said that arthrodesis of the hip joint should not be done because it causes difficulty in sitting comfortably. Here are eight men and women who were just told to sit down. Four have sound arthrodesis of one hip joint and four have not. Can you tell which is which? We doubt whether you can. Certainly all who have looked at this photograph so far have been wrong at least half the time. A patient whose hip joint has been arthrodesed may need to sit slightly forwards on a chair and tuck his foot under it, some chairs being more difficult by reason of cross bars, but to say that there is any real difficulty in sitting comfortably after arthrodesis of the hip is wholly untrue, provided of course that there is not also stiffness of the knee. To say that " high

chairs and stools are almost universally preferred to low soft chairs for sitting " (Stinchfield and Cavallaro 1950) is again entirely different from our experience. These patients can sit in any sort of chair, high or low, hard or soft, if only there is good movement of the knee.

Walking and recreations—There is no patient in the series now reported who after sound fusion of the hip joint needed to use a walking stick. When asked how far they can walk the usual reply is: " as far as I want." This is to be interpreted as anything from three miles to twenty miles according to their instincts and wishes. There is really no limit. It has been said that " walking downhill caused an increase in the severity of the limp and a slowing of pace . . . stair-climbing was awkward " (Stinchfield 1950). This again is exactly contrary to our experience of arthrodesis of the hip. If there is any difficulty at all in walking downhill or uphill, or downstairs or upstairs, the slight stiffness of the hip with free mobility of the knee makes it more difficult to go up than down. But the fact of the matter is that these patients

FIG. 25

In this group there are four people who have had an arthrodesis of the hip joint and four who have not. Without any special instruction they were asked to sit. Can you tell which has an arthrodesed hip joint and which has not? Have a guess.

never wonder which is the more difficult, and there is no question at all of increasing the severity of the limp because they have no limp. Figure 31 shows a patient aged fifty-three years who after arthrodesis of the hip joint has gone back to rock climbing in North Wales and the Lake District. Another patient spends at least one month every winter ski-ing in Switzerland. One is a railway track-man who walks ten miles a day over railway sleepers for five days of each week. One is a porter at Chester station who is no less agile than any other. One is a salesman who drives his car fifteen or twenty miles every day. There are three nurses, and it is interesting to note that one, by reason of the arthrodesis of the hip, was rejected from training in a London teaching hospital but thereafter went to the Robert Jones and Agnes Hunt Orthopaedic Hospital and proved to be one of the best nursing sisters they ever had. There is virtually no occupation or recreation that is impossible after sound fusion of the hip joint in a neutral position—neutral rotation with no more abduction than is needed to correct true shortening, and no more flexion than arises when the patient lies flat on the table.

FIG. 26

FIG. 27

FIG. 28

FIG. 29

FIG. 30

These five patients are a fair sample, representing the degree of knee joint movement that is regained after arthrodesis of the hip joint with immobilisation in plaster for not less than four months. Flexion of the knee joint to the right angle, or even the full range of movement, heel touching buttock, is regained in over 90 per cent of cases.

Pain in the low back after arthrodesis of the hip—Of the 141 patients who were re-examined five to twenty-five years after arthrodesis, ninety declared firmly that they had no pain at all in the low back. Only fifty-one said that sometimes they had pain or discomfort. Even of these, one said that there had never been pain in the back until recent weeks when she had influenza—there are of course many sources of low back pain. Far from it being true that when the hip is fused " excessive strain is thrown on the lumbar spine " or that low back pain is " present in all age groups " we find that 74 per cent of patients had no such pain at all. Perhaps this was because their hip joints were fused in a neutral position with neither excessive abduction nor too much flexion. There is only one patient in the whole of this series who thinks that the pain in the back is worse after the hip joint was arthrodesed. Many said that their low back pain had been relieved or cured by the operation. There can be little doubt that low back pain associated with osteoarthritis of the hip joint arises from unsound ankylosis of the joint in a position of deformity, and from strain of the lumbar intervertebral joints, rather than from increased mobility in consequence of stiffness of the hip. It is time that we renounced once and for all the old view that fusion of the hip causes pain in the low back.

Failure of fusion of the joint after intra-articular arthrodesis—Of the 120 patients with osteoarthritis of the hip joint treated by intra-articular arthrodesis with nail fixation and bone grafting there was failure of fusion in seven. Of these, six had been treated by the former two-stage operation, and one by the one-stage operation. On looking back through the records it seems evident that these failures arose because the patient was released too soon from the full length plaster spica. Even although the hip had been immobilised for several months, undue reliance had been placed on the nail and mobilisation had been encouraged too quickly.

It is true that some of these patients gained considerable relief even from the fibrous ankylosis, but the result in terms of functional activity fell far short of that to be expected by sound bony ankylosis. As soon as it is recognised that fusion of the joint is unsound it is wise to suggest a second operation with further grafting of bone. It is of course hard when a patient has submitted to a big operation and several months of immobilisation in plaster to suggest yet another operation and again another four months in plaster. We can say only that the last patient whose attempted fusion of the hip joint six years ago failed, with pseudarthrosis which in the end caused fracture of the nail, said last week how glad she was to have been persuaded to have another operation. Now that she knows the benefit of a soundly fused hip she thinks that it has all been worth while; and she is sixty-three years of age.

Arthrodesis for congenital subluxation and dislocation of the hip—No hip joint is more easy to arthrodese than a congenital subluxation, the forward and upward displacement of the femoral head making exposure so easy. But when there is complete congenital dislocation of the hip joint, the femoral head being in contact only with the dorsum of the ilium well above the acetabular level, difficulties arise. First, at this level, there is limited contact of the femoral head with the ilium. The ilium against which the femoral head lies is very shallow; anteversion of the femoral neck and other disparities of shape may make it difficult to secure arthrodesis with good apposition of the femur to the pelvis with at the same time correction of true shortening by sufficient abduction of the limb. It may even be necessary to do a subtrochanteric osteotomy at the time of arthrodesis in order to realign the limb. There were eleven patients in this series with complete congenital dislocation of one hip joint all of whom gained sound fusion.

Arthrodesis of the hip for failed cup arthroplasty—In four patients the hip joint was arthrodesed after failed cup-arthroplasty. There is really no difficulty. All that is needed is to remove the vitallium cup, refreshen the joint surfaces, and proceed exactly as for any other arthrodesis.

Arthrodesis for failed McMurray osteotomy—This is a much more difficult problem because there is need to deal both with an arthritic hip joint in which there is still painful movement, and with an ununited subtrochanteric osteotomy. There were four such cases in this series.

Sound fusion was gained in all four, but only with difficulty. It is obvious that the standard operation cannot be performed. The hip joint cannot be dislocated in the ordinary way for full exposure of the acetabulum. All that can be done is to denude the articular cartilage from the roof of the acetabulum and upper part of the head of the femur as far as possible without displacement of the joint. This, together with the impaction of cancellous bone fragments and the insertion of a nail, completes the first part of the operation. There still remains the problem of fusion of the ununited subtrochanteric osteotomy which is best achieved by the grafting of cancellous bone from the ilium rather than by the use of heavy cortical grafts from the tibia (Figs. 34 and 35).

FIG. 31
This man, climbing Gimmer Crag in the Lake District, had an arthrodesis of the hip which is shown in Figures 18 and 19.

CONCLUSIONS AND SUMMARY

1. This is a simple clinical study of the end-results of arthrodesis of the hip joint in patients followed up and re-examined five to twenty-five years after operation.
2. The study was stimulated by our astonishment at recent reports which suggested that arthrodesis of the hip caused serious operative mortality, a high rate of wound infection, and failure of sound fusion in one of every two cases; and that even when sound fusion was gained there was always pain in the back and usually stiffness of the knee. To say that we were astonished puts it mildly.

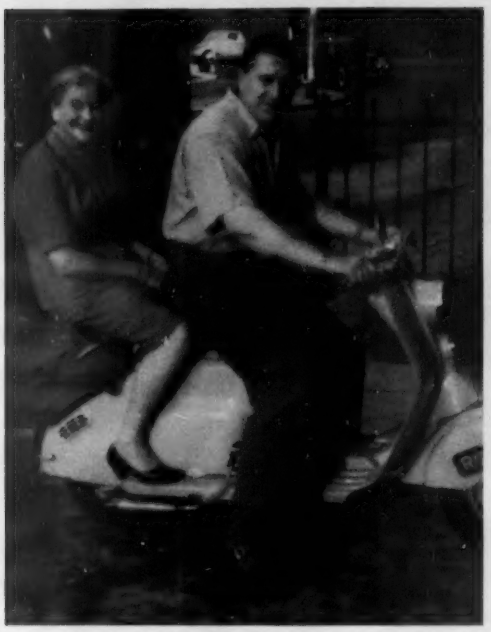


FIG. 32
Do you know which of these two sitting on the motor scooter has had an arthrodesis of the hip? In fact it is the pillion rider.

FIG. 33
This is the patient whose radiographs were shown in Figures 16 and 17. She rides horseback very happily.

FIG. 34
Osteoarthritis of the hip treated by a McMurray osteotomy which failed to unite.

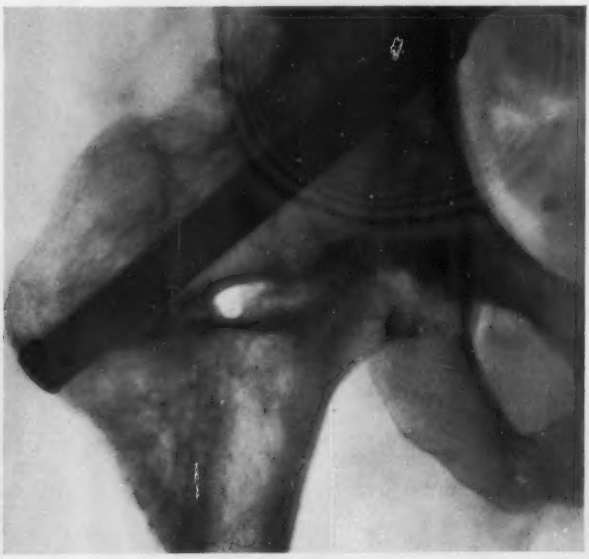


FIG. 35
The hip joint was fused by refreshening the joint surfaces and inserting a nail, the osteotomy being refreshened with grafts of cancellous bone from the ilium. This was one of three ununited McMurray osteotomies for osteoarthritis of the hip in which sound fusion of both hip joint and osteotomy was gained.

3. This review includes 120 patients aged from ten to seventy years, treated for osteoarthritis of the hip joint by intra-articular arthrodesis with the internal fixation of a nail, usually with an iliac graft, and with immobilisation in plaster for not less than four months.
4. Of these 120 patients there was sound fusion of the joint, proved radiographically, in 94 per cent; a mortality of nil; and recovery of free movement of the knee joint to the right angle or far beyond in 91·5 per cent. Almost half of the patients regained normal movement, the heel touching the buttock. Only in eight patients was there less than right-angled flexion.
5. There was no pain in the back—none whatever—in 64 per cent of the patients. In 36 per cent there was some pain or discomfort. One alone said that the low back pain was worse than before the operation. Many others said that pain in the back had been relieved by the operation.
6. It is emphasised that these results were gained only from sound fixation of the joint in the mid-position with neutral rotation, no more abduction than is needed to correct true shortening, and no more flexion of the joint than that with which the patient lies on the table. The limb was immobilised in plaster for at least four months after operation. The stiff knee was mobilised by the patient's own exercise without passive stretching, force or manipulation.
7. Two other groups of patients are considered. There are fourteen treated by fixation of the joint with nail alone, an operation that was never intended to arthrodese the joint and which has long since been abandoned. The other small group is that of patients with old unreduced traumatic dislocation of the hip, a procedure in which the risks of operation are so great and the number of successful results so small as to dissuade us from attempting operative reduction.
8. After successful arthrodesis of the hip joint patients can return to every household activity and every recreation including ski-ing, mountaineering, rock climbing, or whatever else they want.

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FIBROGENESIS IMPERFECTA OSSIUM

A generalised disease of bone characterised by defective formation of the collagen fibres of the bone matrix

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The two cases described here are of special interest because they illustrate a type of skeletal defect not previously known to occur, namely a widespread abnormality in the organic matrix of the bone characterised by a deficiency in collagen fibres. In neither case is there any evidence of a similar defect in the soft tissue collagen. The histological findings were briefly reported by Baker and Turnbull (1950); a more detailed account is given here.

Although apparently never reported before, it is possible that the condition has been encountered by other pathologists, for it would naturally be classed as osteomalacia histologically on account of the large amount of non-calcified bone matrix (osteoid tissue) present. But there are many features—histological, radiological and biochemical—which separate it quite clearly from osteomalacia, and there can be little doubt that the deficient calcification is dependent on the defect in the bone matrix and is unrelated to osteomalacia.

Although this description of the condition is based on only two cases, these show the same histological and radiographic features and differ only in severity, Case 2 being rather more advanced than Case 1.

Both patients were elderly (fifty-six and sixty-four years) and it is clear that the condition was not present early in life but developed, probably within some years of death, by a gradual but widespread replacement of the normal lamellar bone of the skeleton by abnormal fibre-deficient bone.

Since, as will be shown, the essential abnormality arises from a defect in the formation of the fibres of the bone matrix I have named the condition " fibrogenesis imperfecta ossium."

CLINICAL AND RADIOLOGICAL SUMMARY

Only a brief statement of the clinical and radiological aspects will be given here, as these will be dealt with more fully elsewhere.

Both patients were elderly. The first, under Dr Fergus R. Ferguson, was a spinster of fifty-six years; the second, under Dr Donald Hunter, was a man of sixty-four. The clinical features in both consisted of a vague illness lasting some years in Case 1, and for at least three and a half years in Case 2, with the ultimate development of pains in the limbs and trunk. The patients became bedridden with extensive bone changes and spontaneous fractures. No lesion of the nervous system was found. There was no appreciable anaemia, nor any blood cell abnormality. In Case 1 the patient developed xanthomata on the skin shortly before death, but was uncooperative so that no laboratory tests other than a blood count were possible. In Case 2 metabolic studies by Dr Donald Hunter showed that the serum calcium and plasma phosphorus were within the limits of normal, but there was a high alkaline phosphatase in the blood and a high calcium excretion with an unusually high ratio of faecal to urinary calcium—unlike osteomalacia. Radiographic changes were widespread in the skeletons of both cases and included spontaneous fractures of the ribs (multiple in Case 2) and femoral necks (Case 1). The radiographs of the skeleton in Case 1 were described in a report by an experienced radiologist (the late Dr E. W. Twining) as showing a "spotty atrophy," considered at the time to be probably due to widespread tumour deposits. This peculiar spotty appearance, which can be seen in most of the cancellous bone visible in the radiographs of both cases, can be correlated with radiographs of the specimens of femur head and rib from Case 1 and thus with the histological changes.

POST-MORTEM FINDINGS AND NAKED-EYE FEATURES OF THE BONES

Case 1.

Post-mortem findings (summary of findings received from Dr F. R. Ferguson)—The skin showed a xanthomatous patch (5×2 inches) in front of the neck, three xanthomatous nodules on the chest and one in the left groin. The left lung was consolidated at the base and there were pleural adhesions on the right with a healed calcified tuberculous focus at the right apex. Uterine fibroids were present. There was a calcified nodule in the falx cerebri and atheroma of the basilar artery. The brain itself showed no abnormality. The skeleton showed spontaneous fractures of both femoral necks and one rib.

Bone specimens—The following were received (in formol solution): head and neck of femur (neck of femur not examined histologically; lost after war damage); a piece of rib 11 centimetres long including 1 centimetre of costal cartilage; a triangular piece of the vault of the skull 5·0×4·5×4·5 centimetres and 0·5–0·7 centimetre thick.

Femur head—The head, which was almost detached from the neck through a line of tissue judged to be an imperfectly healed fracture, was compressible and cut very easily with a fine saw. The cut surface presented a pale granular appearance; it showed no signs of the normal architecture, and had not the firmness of normal cancellous bone.

Femur neck—The neck had firm cortical bone and the medulla contained fatty marrow.

Rib—This showed no external abnormality, and the cortex was firm. On exposure of the marrow cavity numerous pale chalky nodules, 2–5 millimetres in diameter, were seen to fill much of the marrow space which, however, contained some cancellous bone and some red marrow.

Skull—This was composed of hard bone with a close meshwork of firm bony trabeculae connecting the two tables. No visible abnormality.

Case 2.

Post-mortem findings (by Dr Janet Vaughan, April 1932)—The chief findings were: coronary atheroma; cardiovascular hypertrophy; chronic fibro-caseous tuberculosis of the right kidney and left epididymis; healed tuberculosis of both lung apices; severe osteomalacia (?).

Skeleton—All bones were extremely brittle; there was a fracture of the shaft of the left humerus and one fracture of the shaft of the right femur (probably caused by moving the body). The bones were easier to cut than wood, the broken edges of the fractured bones having a ragged, moth-eaten appearance. There was no deformity of the vertebral column. Diffluent red marrow was present in the centre of the bodies of the lumbar vertebrae, which showed coarse bony trabeculae of peculiar chalky white appearance. Similar appearances were seen on the cut surface of the sternum. The vertebral ends of the lower two ribs on each side showed fusiform swellings, most marked on the left; the soft tissues were adherent to these swellings. The knee joints appeared healthy, with no adhesions or thickening of their synovia. The man was extremely pale and thin, but there was no evidence of anaemia in the organs.

Left femur—The bone showed several post-mortem fractures. It was rather deformed, the shaft being increased in all its diameters and rounded in cross-section with slight anterior bowing and with the condyles turned sharply backwards. The articular cartilages were smooth. The bone cut more easily than wood with the band saw. The shaft of the cut bone could be easily compressed with the fingers, being about 0·6 centimetre thick and porotic. The greater trochanter was also easily compressible and could be cut easily with a knife. In most places the cut surface of the bone showed minute grey granules or groups of granules, which projected from a gelatinous yellow or pink marrow. When isolated the granules were partly gritty and partly tough and elastic. In many places chalky nodules were visible. The cut surface of the outer condyle, for instance, showed brownish-grey and chalky nodules from 0·5–1·0 millimetre in diameter closely packed in a yellow gelatinous marrow. The medullary cavity showed some scattered patches of red marrow in its upper third; apart from this it was filled with fatty marrow.

Ribs—The three lower ribs each showed two fusiform expansions (healing fractures). The corticalis between these expansions was thin, ranging from 1 millimetre to paper thickness, but it was not compressible by the fingers. The cut surfaces showed the grey or yellowish-grey granules described above; red marrow with very little bone was present in a zone 1·5–2 centimetres deep abutting on the costal cartilage.

Lumbar vertebrae—The bodies cut easily with a heavy knife, showing no visible corticalis, and there was deep red marrow with some collections of yellowish-grey granules and a few stellate yellowish-grey or chalky white trabeculae.

Sternum—The cut surface showed three spaces containing mainly red marrow; elsewhere the bone showed the appearance of closely set brownish-white and chalky white coarse granules.

At the necropsy the cut surface of the bones had in general a chalky appearance, which was less obvious when examined after a day in formaldehyde solution.

SUMMARY OF NAKED-EYE FEATURES OF BONES

Naked-eye examination showed, therefore, the following main features of the bones of these two cases. A partial or a virtually complete loss of firm bone, which was replaced by a tissue, granular in appearance, partly tough and elastic and partly gritty in consistence, and showing scattered chalky nodules or coarse bony trabeculae of peculiar chalky white appearance.

The amount of replacement of the firm bone varied at different sites. In some regions, for example the femoral heads of both cases, the whole of the bone appeared abnormal, was lacking in firmness and easily compressible. At other sites, for instance the femoral shaft and ribs in Case 2, much of the tissue was abnormal but some firm cortical bone remained; while in the specimen from the skull in Case 1 the bone did not appear abnormal and was firm throughout. Many of the bones were extremely fragile, as evidenced by spontaneous fractures and crumpling during life and by the occurrence of post-mortem fractures in Case 2.

HISTOLOGY

FIBRE SYSTEMS OF NORMAL BONE AND METHODS OF SHOWING THESE

Since the following account is largely concerned with defects in the fibre systems of the bone matrix it will be as well to preface it with a brief description of the fibre structure of the recognised types of bone tissue.

All bone tissue consists of an organic matrix which calcifies soon after it is deposited, so that even at sites of active bone formation only a small amount of uncalcified organic matrix (osteoid tissue) is present; at sites where there is no sign of osteoblastic activity no uncalcified matrix is normally found.

The organic matrix is composed largely of collagen fibres which are embedded in an amorphous " cement substance " or " ground substance."

Note—The organic matrix is composed largely of the fibrous protein collagen together with a polysaccharide (or polysaccharide-protein complex) which probably constitutes the amorphous " ground substance." The ground substance is not birefringent with polarised light and appears as a non-staining material between the fibres when these are stained by silver methods or by Schmorl's Thionin.

Lamellar bone, which constitutes nearly the whole of the post-infantile skeleton, owes its lamellated appearance to the arrangement of its fibres. These are very fine and closely packed and are arranged in sheets. In each sheet, forming a single lamella, commonly 2–5μ thick, the general direction of the fibres is the same, but in the next sheet the direction has changed (like the grain of plywood). This alteration in fibre direction produces the lamellation which is most distinct where the change of angle is maximal (90 degrees) but is quite obvious where the change is through 45 degrees, which is the average figure. Various grades of lamellation occur, and a non-lamellar or imperfectly lamellar bone made up of sheets of fine fibres but with little or no alteration of fibre direction occurs normally in young children, particularly in more recently formed bone near the epiphyses, where most of the fibres run parallel to the longitudinal axis of the trabeculae.

Non-lamellar bone—In the most common type of non-lamellar bone the fibres of the matrix cross one another at all angles like an irregular basketwork or feltwork. Usually the fibres vary considerably in thickness, some being very coarse.

Note—There is no standard nomenclature for this type of bone. Koelliker used the term " Faserknochen " or fibre bone. The term coarse-fibred bone is also used, and some writers (incorrectly) call it metaplastic bone. v. Ebner (1875) called it " geflechtartiger Knochen," a term which has been widely used in the German literature, and H. M. Turnbull for many years used an equivalent English term " woven bone."

" Woven bone " seems the best term for it, as this emphasises the characteristic woven feltwork of fibres.

The usual type of woven bone is coarse-fibred, but less commonly it is composed only of fine fibres—" fine-fibred woven bone."

The primitive bone found in the embryo is the coarse-fibred woven type which is gradually replaced by lamellar bone as development proceeds; but it reappears in all sorts of pathological conditions where new bone is rapidly formed, for example in fracture callus. It may form on a surface of pre-existing bone, but often arises in a mass of reactive fibrous tissue, where it commonly incorporates many of the pre-existing fibres; new fibres and also cement substance are laid down around these to produce the woven-bone matrix.

Lamellar bone with its more highly organised fibre systems does not form as rapidly as woven bone, and can form only on a firm surface which is nearly always pre-existing bone (lamellar or woven) or calcified cartilage. It is usually deposited on the surface of woven-bone trabeculae formed during the earlier more active stage of a pathological process, thus producing lamellar bone trabeculae with central cores of woven bone. The reverse process also occurs, residual fragments of lamellar bone, the bulk of which has been removed by osteoclasis, being incorporated in a new deposit of either lamellar or woven bone. Such fragments have sharply defined margins pitted by Howship's lacunae (" lacunar margins ")—evidence of the earlier osteoclasis of their surfaces. As will be seen later this is a frequent finding in the specimens of the two cases here reported; fragments of lamellar bone with lamellar margins being incorporated in trabeculae of abnormal fibre-deficient bone.

Metaplastic bone—This term should be used only for bone produced by ossification of a mature non-bony tissue, that is of bone in which all or nearly all the collagen fibres of the matrix are those of a pre-existing collagenous structure. Such bone is normally present in small amounts on the subperiosteal surfaces of lamellar bone in many regions, particularly where fasciae, ligaments or tendons pass into the periosteum. It results from ossification of fibre bundles in the deepest part of the periosteum. Such bundles may run parallel to the surface, or they may pass into the lamellar bone, where they form Sharpley's fibres.

Such periosteal metaplastic bone can be recognised histologically by the fact that its fibre bundles are continuous with those of the surrounding periosteal fibres, as can be seen by the use of polarised light.

In several of the bone samples from Case 2 abnormal fibre-deficient bone has replaced the whole of the normal cortex with the exception of a thin layer of metaplastic bone on its surface.

METHODS FOR SHOWING THE FIBRE STRUCTURE OF BONE

Owing to the fact that the fibres and the cement substance in which they lie have virtually the same refractive index, the fibre structure is not visible in bone sections stained by the usual routine methods. The following methods for showing it have been used in this study.

Schmorl's Thionin method for cell spaces, canaliculi and fibre systems—This is satisfactory only with celloiden sections, with which it can give beautiful pictures, but it is difficult and erratic in its results. It does not show the fine fibrillary structure of the abnormal bone and osteoid tissue in these cases.

Polarised light—von Ebner (1874) showed that sections of bone were birefringent with polarised light. This is due to their collagen fibres, which are birefringent. With a microscope fitted with a suitable polarising attachment they shine up against a dark background when correctly orientated in relation to the plane of polarisation.

The method has been used by Petersen (1930) and others for studying bone structure. It has the great advantage of simplicity, because it can be used on routine sections without special staining, and I have used it for many years. Collagen fibres are not highly birefringent, so that a good optical system and a strong light are needed to get good results, and a rotating microscope stage is very desirable. It is very useful for distinguishing different types of bone, and was particularly valuable in these cases because its use as a routine procedure when examining the sections from Case 1 first revealed the deficiency in the fibre content of the bone tissue.

The method has the disadvantage that only those fibres that are correctly orientated can be seen or photographed at one time, and the section must be continually rotated in order to reveal all the fibres; moreover it fails to show up the finer details of fibre structure. The birefringence of collagen fibres is greatly increased in sections prepared by the silver methods described below, but though they are much brighter under the polarising microscope there is further loss of detail.

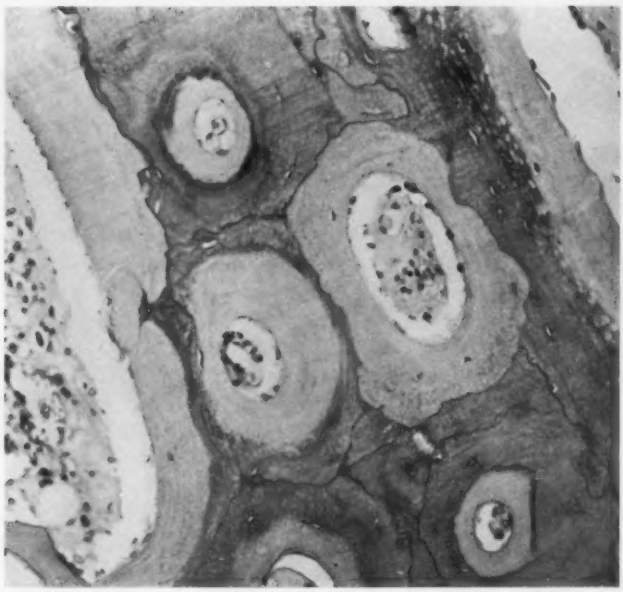

FIG. 1

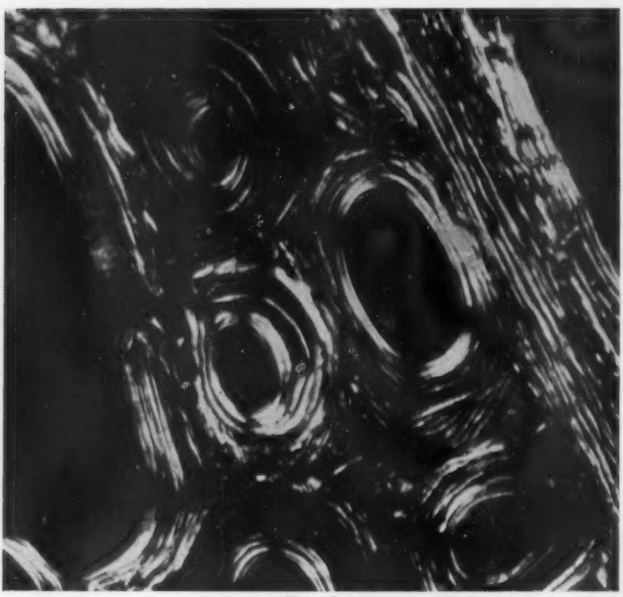

FIG. 2

Osteomalacia showing normal lamellar structure of osteoid tissue.
Figure 1—Cortex of rib showing calcified bone (dark) with wide osteoid borders (pale) on surfaces and filling inner part of Haversian systems (one system entirely osteoid). Figure 2—The same using polarised light, showing lamellar structure with no distinction between osteoid tissue and bone. (Paraffin section. Iron haematoxylin and eosin, ×190.)

Silver methods for fibres—I have found that these methods are best for showing fine detail in the fibre structure of bone, and have for some years used mainly Gömöri's reticulin method with gold toning (Gömöri 1937), the best detail being obtained with thin paraffin sections. The fine detail

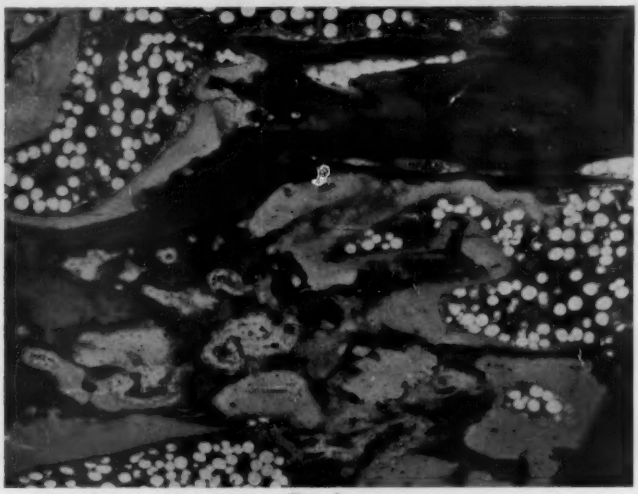

FIG. 3

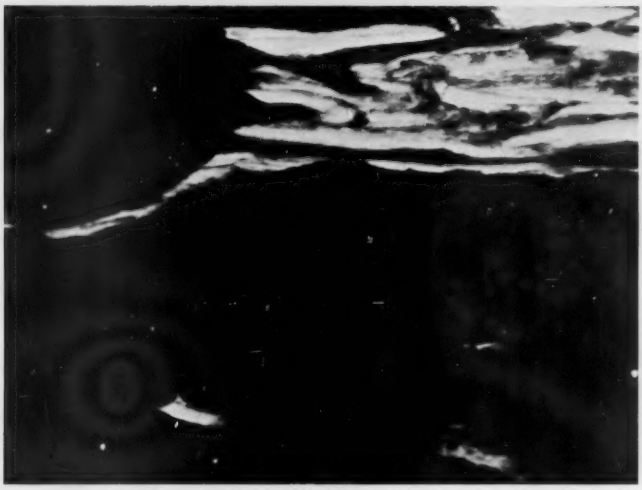

FIG. 4

Showing absence of birefringence in the fibre-deficient bone and osteoid tissue. Field from the right-hand side of Figure 9.

Figure 3 shows calcified bone (dark) and osteoid tissue (light grey) with fatty marrow (white circles = fat cells). Figure 4—The same with polarised light. The lamellar bone in the upper part of the figure is highly birefringent and two small fragments are also seen towards the lower margin of the figure. The rest of the tissue both calcified and non-calcified is fibre-deficient and shows no birefringence. (Staining as in Figure 9, ×33.)

(*Note:* The small bright dots seen in Figure 4 and many of the polarised light pictures are fine particles of dust unavoidably present in many mounted sections.)

of the fibrillary structure of the abnormal bone shown in Figure 33 was obtained with 5μ paraffin sections using Gömöri's method. In satisfactory preparations of decalcified normal bone the fibres are aureophil (reddish brown) with some argyrophil (black) markings produced mainly by the walls

FIG. 5

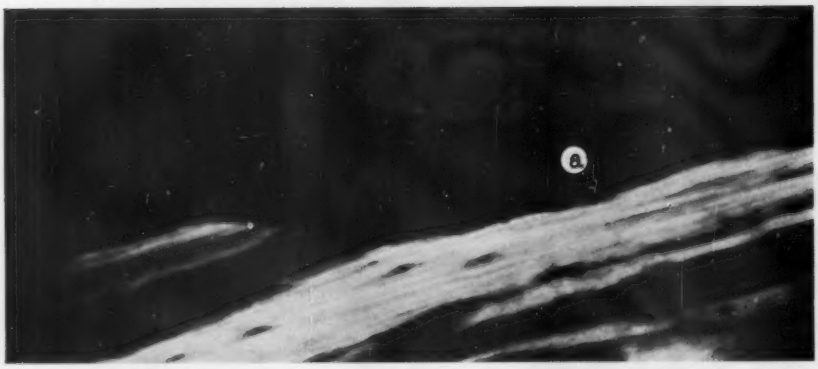

FIG. 6

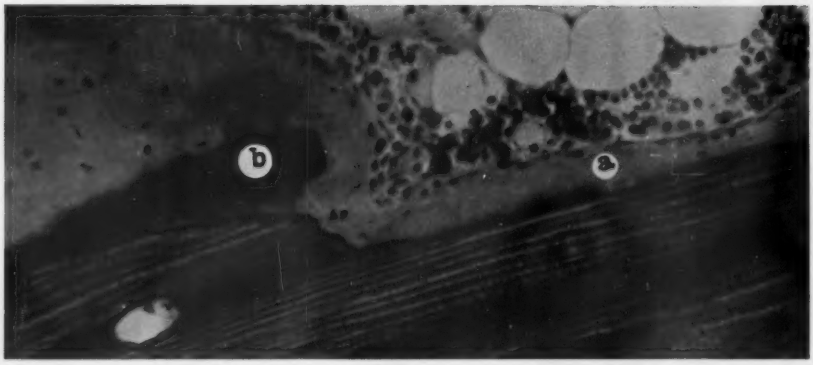

FIG. 7

Showing that the abnormal osteoid tissue is not birefringent and shows no fibre structure with Schmorl's Thionin stain.

Figure 5—Part of Figure 9 showing darkly stained lamellar bone (below) on the upper surface of which a border of abnormal osteoid tissue (*a*) has been deposited. This shows slight granular calcification (patchy clouding) in places, and a denser calcified patch at (*b*). (Ehrlich's haematoxylin and eosin, × 200.) Figure 6—The same using polarised light. Only the lamellar bone is birefringent. Figure 7—The same region in a neighbouring serial section stained by Schmorl's Thionin for fibres and canaliculi. The structure of the lamellar bone is well seen. The abnormal osteoid tissue stains lightly and shows no fibre structure. Cell nuclei in the osteoid border and also the calcified area (*b*) stain more deeply.

of the cell spaces and canaliculi which are argyrophil. In bone sections in which the haematoxyphil staining of areas of granular calcification has been retained (see " calcification ") the fibres in these areas are argyrophil (Fig. 33). A bundle of collagen fibres passing into such an area changes from brown to black as it enters the calcified region. The reason for this is not clear.

OUTLINE OF MAIN HISTOLOGICAL CHANGES SEEN IN HAEMATOXYLIN AND EOSIN STAINED SECTIONS, SHOWING INTERMEDIATE AND SEVERE GRADES OF THE DEFECT

As indicated by the naked-eye appearances, various grades of defect were found in the different bone samples examined.

Example of intermediate grade of the defect—It will be simplest to describe first the microscopic appearances of a region where an intermediate grade of defect is present, the original lamellar bone being only in part replaced by defective bone. Figure 9 shows a field of the porotic femoral cortex of Case 2. The original lamellar bone of the cortex has been hollowed out, and in the spaces so produced a lightly eosinophilic, largely non-calcified tissue has been deposited on the bone surfaces. The new tissue forms relatively thick layers and irregular masses and virtually fills many of the spaces in the old bone. This imperfectly calcified new tissue resembles the osteoid tissue in osteomalacia and the histological picture suggests a very advanced stage of this disease.

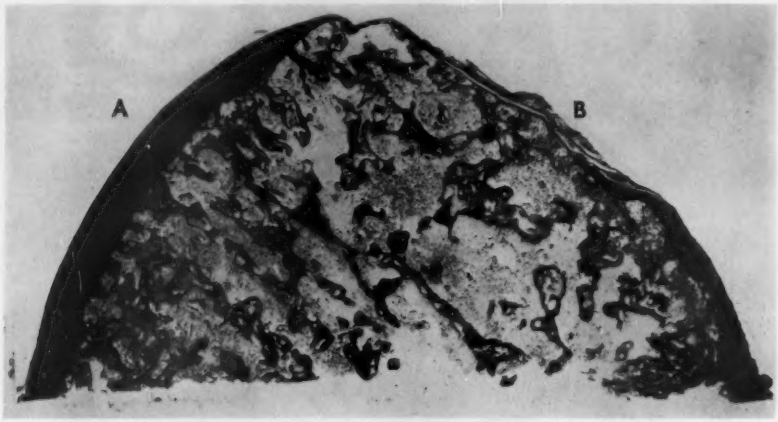

FIG. 8

Severe grade of the defect. Part of lateral condyle of femur, Case 2. Articular cartilage A, lateral surface of condyle B. The cancellous bone shows thick irregular widely spaced trabeculae which are composed largely of osteoid tissue (see Fig. 10). (Whole section, ×2½.)

Further investigation has, however, shown that there is an important difference between this " osteoid " and the osteoid tissue of osteomalacia. In osteomalacia it is well established that, as Pommer (1885) stressed, there is no histological defect in the organic matrix of the bone; this is deposited in the normal manner and shows a normal structure, but fails to calcify owing to extra-skeletal factors—usually vitamin-D deficiency. The normality of the organic matrix is further proved by its rapid calcification with successful vitamin-D therapy. Histologically the osteoid tissue in osteomalacia shows the normal collagen structure with its lamellar pattern, as can be readily demonstrated by the polarising microscope. Figures 1 and 2 demonstrate the normal lamellar structure of the osteoid tissue in osteomalacia. Under polarised light (Fig. 2) the pale osteoid tissue seen in Figure 1 shows the same lamellar structure as the darker (calcified) bone.

The osteoid tissue in the two cases here reported does not show a normal fibre structure—a point first noticed when examining sections of Case 1 with the polarising microscope. It was

FIG. 9

Cortex of femur, Case 2. The cortex is thin (about 3·5 millimetres at this point) and porotic, showing many spaces filled by haemopoietic and fatty marrow. Only part of the bone (blue) is calcified: wide non-calcified (pink) borders of osteoid tissue cover most of the surfaces, and there is an irregular mass of this near the lower right-hand corner of the figure. The medullary cavity (lower third of figure) contains fatty marrow with small collections of haemopoietic cells. [All the osteoid tissue seen here is fibre-deficient. Most of the calcified tissue is lamellar bone, but some is fibre-deficient bone (see Figs. 2 and 3).] (Celloidin section; Ehrlich's haematoxylin and eosin, ×12.)

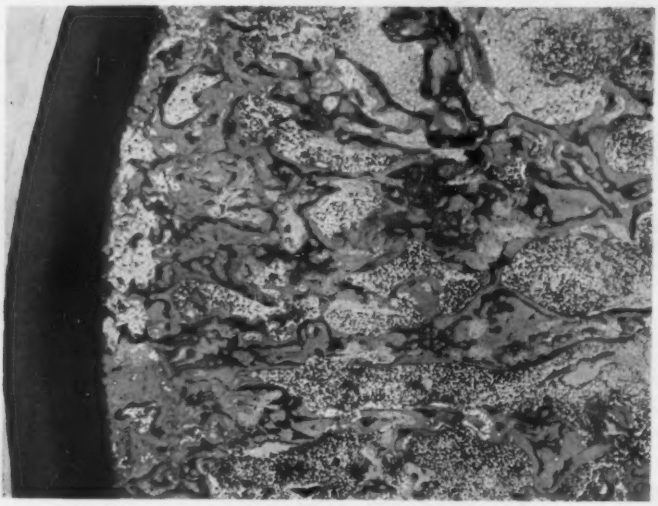

FIG. 10

Showing a severe grade of the defect. Lateral condyle of femur, Case 2. Normal articular cartilage to the left. The cancellous bone shows thick irregular trabeculae composed largely of osteoid tissue (pink) in which are irregular strands and patches of calcification (blue). Fatty marrow with a little haemopoietic tissue fills the cancellous spaces. All the bone tissue, calcified and uncalcified, seen here is fibre-deficient. (Celloidin section; Ehrlich's haematoxylin and eosin, ×7.)

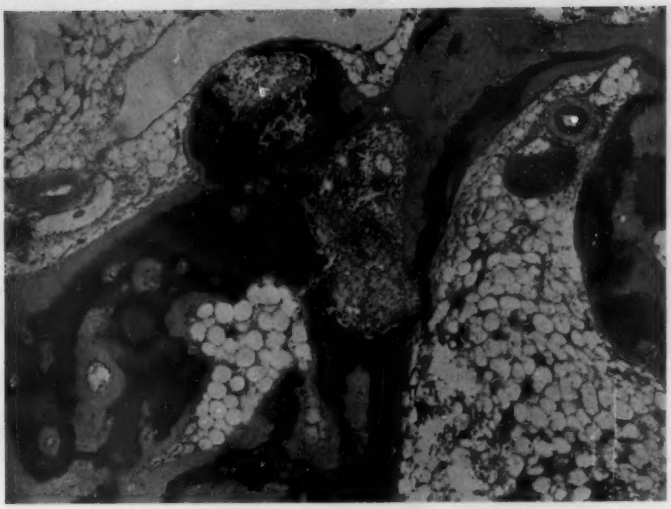

FIG. 11

Foci of osteoclastic absorption on trabeculae of lateral condyle of femur (*Case* 2). Trabeculae showing much osteoid tissue in which are irregular strands and patches of deeply haematoxyphil (dark) calcification. Near the centre of the figure a trabecula has been hollowed out, leaving an oval space filled by fibroblastic tissue and containing small blood vessels. Numerous osteoclasts are present at the periphery of the space, mainly on the surface of the calcified tissue which forms the lower and left-hand walls of the space. To the left of the upper part of this oval space is a similar but smaller space, showing numerous osteoclasts. (Celloidin section; Ehrlich's haematoxylin and eosin, ×45.)

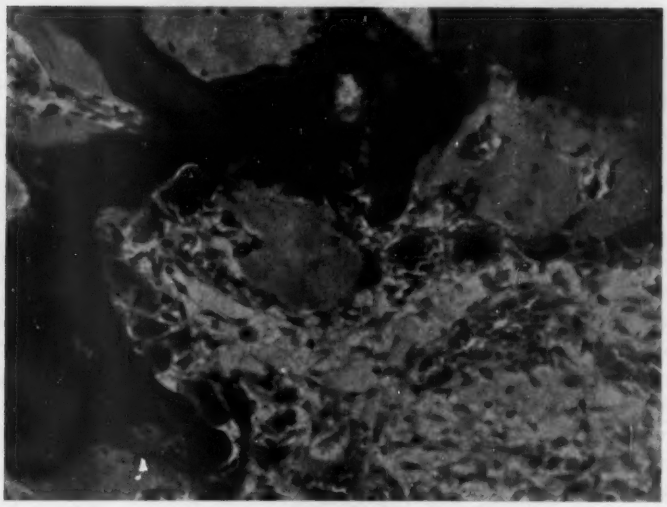

FIG. 12

A similar focus showing osteoclasts in contact with calcified tissue (left): some also applied to the surface of osteoid tissue (centre and right). (Owing to the deep haematoxylin staining of the cytoplasm of the osteoclasts their nuclei do not show up in the figure.) (The same section as Figure 11, ×200.)

seen that the tissue showed no lamellar structure and was only very slightly refractile, with a scarcely visible feltwork of very fine fibres (unlike either lamellar or woven bone).

Figures 3 to 7 demonstrate that the osteoid tissue in these cases is very poorly refractile with polarised light. (No refractility is visible in these photographs, but by direct vision with the polariser under optimal conditions a faint network of fine fibrils can be distinguished.) Nor does it show any fibre structure by Schmorl's Thionin method, but stains a diffuse sky-blue. A marked deficiency in fibres is found in the osteoid tissue of all the material from these two cases and distinguishes it from the osteoid tissue in osteomalacia or rickets.

While much of this abnormal bone matrix remains uncalcified, some of it, mainly the older parts, calcifies producing the irregular deeper blue streaks and patches seen in Figure 9 and the haematoxyphil cores of the trabeculae in the femur head and elsewhere (Fig. 24). The bone resulting from this calcification remains abnormal, being fibre-deficient like the osteoid tissue. Further details of the calcification are described later.

The medullary cavity and marrow spaces in the bone contain a mixture of fatty and haemopoietic marrow (Fig. 9) without fibrosis or other abnormality.

Example of advanced grade of the defect—Where an advanced grade of the defect is present almost the whole of the lamellar bone has been replaced by the abnormal fibre-deficient and only partially calcified bone. This grade of defect was present in the specimen from the femoral heads of both cases and in those from the greater trochanter, the lateral condyle of the femur and body of lumbar vertebra in Case 2.

In most of these specimens there is evidence of crumpling and distortion of abnormally soft trabeculae, which in places form compacted masses with obliteration of cancellous spaces, and there may be fractures or partial fractures which complicate the histological picture. But the section of the lateral condyle of the femur (Case 2) shows an advanced grade of the condition without signs of serious crumpling or reaction to trauma. It is therefore a good example of the advanced but uncomplicated defect. The section is bounded on one side (Fig. 8) by the lateral surface of the condyle and below by the lateral part of the articular cartilage. Most of the section is occupied by cancellous bone composed of thick irregular osteoid trabeculae partially calcified (blue streaks and patches in Fig. 10). Between the trabeculae the wide cancellous spaces contain fatty marrow with a little haemopoiesis. The trabeculae in the central region of the section are directed approximately vertically to the articular cartilage (Fig. 8) and are connected laterally with similar trabeculae supporting the lateral surface of the condyle. They therefore have the normal arrangement in relation to the stresses in this region, but they are much thicker and much less numerous than normal. A comparison with a normal specimen from the same site shows that the calcified strands within the thick osteoid trabeculae are often as wide as, or wider than, the normal trabeculae.

The lateral surface of the condyle is covered by periosteum beneath which is a thin shell (0·05–0·5 millimetre thick) of calcified bone. Attached to the deep surface of this shell is an irregularly trabeculated layer of partly calcified osteoid tissue similar to that described above. The calcified shell is composed of metaplastic bone containing coarse fibres which are continuous with those of the deep layer of the periosteum (as in Figure 27). It contains no lamellar bone. (This region in a normal specimen also shows a very thin outer shell bounding the cancellous bone, but two-thirds or more of its thickness is lamellar bone, one-third or less on the surface being metaplastic bone.)

In this condyle in Case 2 the deep surface of the shell of metaplastic bone presents a serrated appearance, being pitted by innumerable Howship's lacunae (as in Figure 26). This irregular surface is for the most part in contact with an underlying layer of fibre-deficient osteoid tissue which has been deposited on it. In most of the limited areas where the metaplastic bone is in contact with the cellular marrow tissues osteoclasts are present on it.

Apart from the shell of metaplastic bone the only normal bone in the section consists of a few small scattered fragments of lamellar bone, some of them embedded in the osteoid

FIG. 13

A fragment of lamellar bone lying in partly calcified fibre-deficient bone matrix. The same field as shown in Figure 15 (see description of this). For comparison with Figure 14.

FIG. 14

The same using polarised light which shows up the fusiform fragment of birefringent lamellar bone in contrast to the surrounding non-refractile fibre-deficient bone. A second very thin fragment of lamellar bone is seen below.

trabeculae, others attached to the base of the calcified zone of the articular cartilage along the chondro-osseous junction. Using polarised light such fragments show the refractility of normal bone in contrast to the surrounding non-refractile fibre-deficient osteoid tissue or bone.

Active osteoclasis is found not only on the deep surface of the shell of metaplastic bone but also at various sites in the partly calcified osteoid trabeculae where osteoclastic tissue penetrates in places into the trabeculae, producing absorption of parts of both calcified and non-calcified bone matrix. Such a focus of absorption presents a rounded or irregular bay or space filled by vascular spindle-cell tissue in the periphery of which are numerous osteoclasts in Howship's lacunae on the bone surfaces (Figs. 11 and 12).

The articular cartilage appears normal; it stains normally and with polarised light shows the normal diffuse sheen produced by its fine fibrillary structure. This agrees with findings in other sites (*vide* cartilage of femoral heads and ribs).

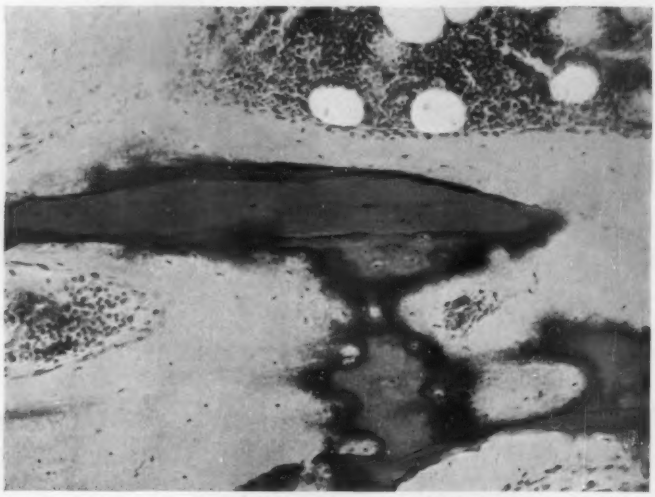

FIG. 15

To show types of calcification. This is part of Figure 9 and shows a fusiform fragment of lamellar bone (defined by polarised light in Figure 14) lying in fibre-deficient bone matrix, seen as poorly staining osteoid tissue which, with two marrow filled spaces (above and lower left) makes up the background of the picture. This osteoid tissue has calcified round the margins of the fusiform fragment, and in irregular patches below it. The fragment is stained a diffuse blue seen in all the calcified lamellar bone in this section. The calcified fibre-deficient matrix shows patches of diffuse blue staining, similar to that of the lamellar bone, but this has wide margins with a deeply haematoxyphil granular appearance characteristic of much of the calcification in the abnormal bone. (Section as Figure 9, ×140.)

CALCIFICATION

Most of the material used for the study of the details of calcification was prepared by slow (three months) decalcification in Müller's fluid followed, usually, by a final treatment with 3 per cent formic acid. Celloidin sections were stained by Ehrlich's haematoxylin (twelve to fifteen hours) and eosin. Very good haematoxylin staining of the calcified parts of the abnormal bone, with, however, poor staining of the lamellar bone, was obtained with formal-fixed material (without Müller treatment) decalcified in formic or nitric acid.

In successful haematoxylin and eosin stained sections produced by the first method calcified lamellar bone stains a diffuse slaty-blue, while any non-calcified (osteoid) borders on its surface are eosinophilic. Calcified coarse-fibred woven bone stains deep blue in the form of deeply staining fine granules, discrete in places where calcification is starting but forming deep blue opaque masses where it is more advanced. In the central (older and more mature)

regions of calcified woven bone trabeculae the staining becomes rather lighter and more diffuse, similar to but darker than that of lamellar bone.

The diffuse blue staining of calcified lamellar bone is easily removed by treatment with acids, and considerable care and judgment are therefore needed to produce sections in which it is satisfactorily retained. The deep blue haematoxyphil staining of the granular calcification of coarse-fibred woven bone shows, on the other hand, considerable resistance to acids and can be retained without much difficulty in formalin-fixed material decalcified in nitric acid, provided the acid treatment is not unnecessarily prolonged.

Examination of a section of normal lamellar bone with osteoid borders, prepared as

FIG. 16

Rib cortex of Case 2. At the top of the figures is some fatty tissue; below this is the periosteum, the fibres of which run horizontally. Immediately beneath the periosteum (central) is a small fragment of normal (metaplastic) bone. The remainder of the cortex (apart from one other small fragment) is abnormal fibre-deficient bone partly calcified to form an irregular trabecular pattern (dark purple) with a large amount of surrounding osteoid tissue (pink). At the bottom of the figure is the haemopoietic marrow of the medulla (see also Figs. 17 and 18).

described above, shows at the junction of the eosinophilic osteoid border with the diffusely haematoxyphil calcified bone a thin deeper blue line. Under high magnification this line appears finely granular, like the calcification in woven bone. This is the region where the osteoid tissue is at the early stage of calcification. Since during the growth of lamellar bone new layers of osteoid tissue are being continually deposited on the growing surfaces and are calcifying in their deepest part (so that the osteoid borders do not at any time exceed about 7–10μ in thickness in normal growing bone) it is clear that the thin line of early granular calcification, which does not increase in width, changes into the non-granular, less intensely haematoxyphil state. This is presumably the result of a finer dispersion of the calcium salts with the ultimate development of a homogeneous crystalline structure.

A similar change occurs in the granular calcification of woven bone but is longer delayed and is less complete. Calcifying cartilage also shows the same phenomenon, passing from a granular to a diffuse type of calcification.

The factors responsible for the haematoxyphil staining of calcified bone are not clearly understood. It is clear that calcified bone will still stain blue with haematoxylin after mild treatment with acid and/or Müller's fluid which has been sufficient to render it negative with the Kossa reaction for calcium

FIG. 17

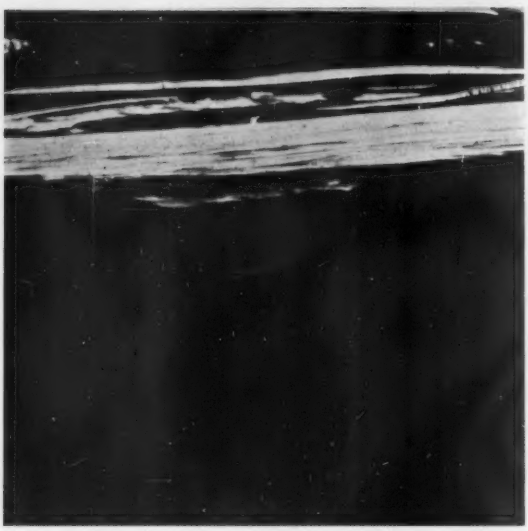

FIG. 18

Showing periosteum with normal fibre content covering fibre-deficient bone. The same field of rib cortex shown in Figure 16 viewed by normal and polarised light. The abundant collagen fibres of the periosteum are highly birefringent (Fig. 18) whereas the underlying bone shows (apart from two small fragments of normal bone) no birefringence. (Section as Figure 16.)

(strictly speaking for phosphate (Cameron 1930)). Some haematoxyphil substance (rather more resistant to acids and Müller's fluid than is the calcium salt) must therefore be produced in the bone matrix at sites of calcification. Though there have been speculations as to its nature nothing definite is known. It may be a mucopolysaccharide (Rubin and Howard 1950).

Haematoxylin used with due precautions is much better for defining details of calcification than is the Kossa reaction, which produces a black mass blotting out all structure.

Four types of calcified tissue can be recognised in sections from these two cases: 1) normal calcified lamellar bone showing the diffuse light blue haematoxylin staining; 2) coarse-fibred woven bone (in relation to the fracture of ribs in Case 2) showing the characteristic strongly haematoxyphil granular calcification described above; 3) abnormal fibre-deficient bone matrix showing diffuse non-granular blue staining similar to but usually rather darker than that of the lamellar bone; and 4) abnormal matrix showing deeply haematoxyphil granular calcification like that of woven bone. Examples of (1), (3) and (4) are seen in Figure 15, showing a fragment of lamellar bone surrounded by diffusely calcified abnormal matrix with dark-staining granular calcification at its edge and extending as clouds of granules into the surrounding abnormal osteoid tissue. Figure 14 is the same field under polarised light, showing

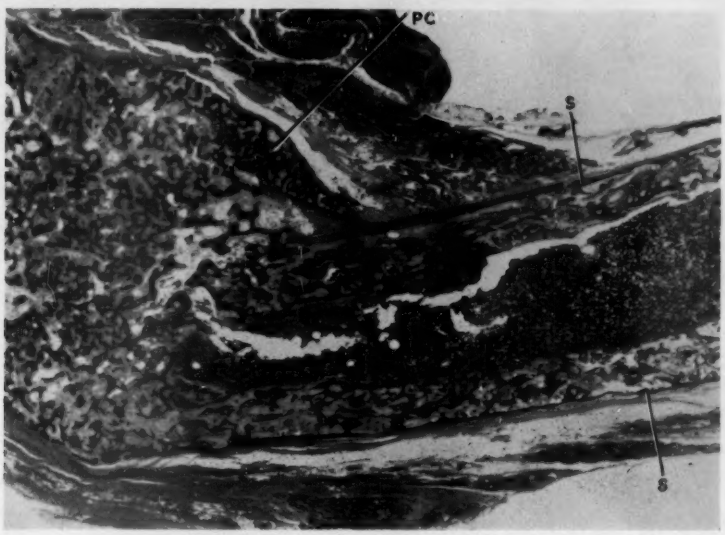

FIG. 19

Angulated healing fracture of rib (*Case* 2). A mass of callus filling the medulla and replacing the cortex is seen on the left of the figure. To the right is undisturbed cortex and medulla which is filled with haemopoietic marrow. The cortex is composed of partly calcified fibre-deficient bone (as seen in Fig. 16) except for a very thin outer shell of metaplastic bone (fine, dark subperiosteal line S, S). Nearly the whole of the callus mass is also fibre-deficient: the chief exception being the periosteal callus (PC) which consists of a layer of irregular trabeculae of coarse-fibred woven bone. (Celloidin section; Ehrlich's haematoxylin and eosin, ×6.)

the double refractile collagenous structure of the piece of lamellar bone in striking contrast to the poorly collagenous non-refractile matrix of the abnormal bone and osteoid tissue. The areas of diffuse non-granular calcification in the abnormal matrix clearly represent the fully calcified state of this tissue—the nearest it can get, so to speak, to normal bone—and it is difficult to distinguish some of the areas of this tissue from normal bone in haematoxylin and eosin sections without using the polariser.

The granular calcification is the earlier, less complete, stage of the process; and much of the abnormal matrix either fails to calcify or shows the granular type of calcification.

The relatively intense haematoxyphil staining of the calcified abnormal bone enables it to be easily identified in the Haversian systems of the skull and ribs of Case 1, as shown below.

About half, or at some sites more than half, the abnormal tissue is entirely uncalcified, forming the large amount of lightly eosinophilic fibre-deficient osteoid tissue which is the

outstanding feature in sections stained with haematoxylin and eosin (Fig. 10). The calcified areas are mostly the older parts of the tissue forming the central parts (cores) of osteoid trabeculae. These calcified cores are made up of a mosaic of fragments of different ages, many with deeply pitted borders as a result of earlier osteoclastic erosion. In the central (axial) parts of the cores the calcification is usually less heavily haematoxyphil and more diffuse than in these outer, more recently calcified parts which show dark blue granular calcification extending with an ill-defined edge of scattered granules into the osteoid zone (Fig. 24).

These features correspond to those seen in osteomalacia, but with an important difference. In osteomalacia the calcified cores of trabeculae are composed of a mosaic of fragments of calcified lamellar bone dating from the pre-malacic period. In addition, a limited amount of granular calcification occurs (as noted originally by Pommer) in the osteoid tissue, particularly in the deeper part abutting on the calcified cores—indicating that, even in the course of a severe osteomalacia, a small amount of incomplete calcification occurs in the osteoid tissue. In this material, on the other hand, the calcified cores of the trabeculae are composed not of the original lamellar bone (except as a few isolated fragments) but of the older parts of the fibre-deficient matrix which have calcified during the course of the disease.

In osteomalacia, since the malacic bone is deficient only in calcium salts, it can revert to histologically normal bone if calcification is promoted by successful therapy. In fibrogenesis imperfecta, on the other hand, although part of the abnormal bone calcified spontaneously during the disease, the calcified portions still remain grossly abnormal histologically (and also, no doubt, mechanically) owing to their lack of collagen fibres.

The coarsely granular form of the calcium salt deposit in much of the abnormal bone appears to be the cause of the chalky appearance noticed in the bones in Case 2 at the necropsy, and very obvious in the chalky nodules seen in the rib in Case 1. Thin pieces of normal bone, though opaque when dried, are translucent in the moist state. The translucence is no doubt due to the fine dispersion of the calcium salt in the organic matrix. Coarse granules would reflect light, producing an opaque appearance; and the lack of collagen, with resulting diminished refractility of the matrix, probably plays a part also.

APPEARANCES AT OTHER SITES SHOWING INTERMEDIATE AND SEVERE GRADES OF THE DEFECT

Ribs in Case 2—In the parts of two lower ribs examined histologically the picture is complicated by the presence of four healing fractures, two in each rib. An undamaged length of each rib including part of the costal cartilage was, however, available, and both ribs show essentially the same features. The periosteum shows no abnormality and contains many well formed, strongly birefringent coarse fibre bundles running parallel to the long axis of the rib (Figs. 17 and 18). The cortex, which is as thick as, or rather thicker than, normal (1–1·3 millimetres), is composed almost entirely of fibre-deficient bone and osteoid tissue. The bone forms an irregular trabecular pattern, with wide osteoid margins and patches of osteoid tissue (Fig. 16). The only bone with a normal fibre content, apart from a few isolated fragments of lamellar bone, is a very thin outer shell (0·02–0·2 millimetre thick) attached to the deep surface of the periosteum. This shell is composed almost entirely of metaplastic bone produced by ossification of the coarse parallel fibre bundles of the deepest part of the periosteum or in some regions of finer oblique fibres. The medullary cavity contains a mixture of haemopoietic and fatty marrow, with only a few fine trabeculae of fibre-deficient bone with osteoid margins.

Abnormal bone and osteoid tissue like that forming the rib cortex bridges the marrow cavity at the level of the chondro-osseous junction and extends up inside the costal cartilages, which have been eroded centrally. A few scattered fragments of lamellar bone with lacunar margins are found embedded in this abnormal bone, showing that the central part of the cartilage was first ossified by normal bone replacement, this being again replaced by abnormal bone.

Rib fractures—One rib has two fractures, one anteriorly, the other involving the head. The anterior fracture is angulated with a mass of bony callus situated about 2 centimetres from the chondro-osseous junction. The callus shows a mass of irregular trabeculae filling the medulla and replacing the cortex. The medullary trabeculae are composed of fibre-deficient bone and osteoid, but the bone developed in the thickened periosteal tissue forms a layer of woven-bone trabeculae containing many coarse fibres running in from the periosteal surface. This layer is very thin on the convexity, but thicker on the concavity of the angulation, where it extends along the shaft of the rib for a short distance between the cortical shell of metaplastic bone and the periosteum (see Fig. 19).

While, therefore, the internal callus is composed of fibre-deficient bone, the subperiosteal callus is similar to that commonly found in fractures of normal bones.

In the substance of the periosteum to one side of the fracture line is a small lenticular abscess

FIG. 20

FIG. 21

Healing rib fracture (Case 2) without displacement; to show fibre production in periosteal callus but very little in the deeper callus. Figure 20 shows a small mass of periosteal callus with deeply stained well calcified trabeculae of coarse-fibred woven bone. This is situated above the line of the cortex: the arrow marks a layer of normal metaplastic bone which formed the outer shell of the cortex. Figure 21—The same with polarised lighting, showing birefringence of periosteum with much formation of birefringent fibres in the periosteal callus (arrow as in Figure 20), but very little in the deeper callus. (Celloidin section; Ehrlich's haematoxylin and eosin, ×18.)

(2×6 millimetres) containing a minute fragment (1×0·5 millimetre) composed of fibre-deficient bone and osteoid tissue. This was the only inflammatory lesion encountered in the bone material of these two cases. It is clearly not of long standing.

The head of this rib shows a crumpled, elongated mass of fibre-deficient spongy bone and osteoid, representing the expanded head with the narrow neck recognisable at one end. A very thin outer shell of normal (mainly metaplastic) bone covers the neck and extends as an incomplete shell over the mass. A band of small closely packed trabeculae about 3 millimetres wide extends across the middle of the mass and marks the line of a fracture. This line of callus is composed mainly of fibre-deficient bone and osteoid, but at each end, where the periosteum is involved, a subperiosteal layer of coarse-fibred woven bone, similar to that seen in the anterior fracture site, is present.

The two fractures in the other rib are in the shaft, about a centimetre apart. The essential features are the same as in the two just described, but there has been no alteration in the line of the rib shaft, and the amount of callus is less. Subperiosteal woven bone is present over the fracture line, and at one point is a small mass of fibrocartilage. Most of the deeper callus is fibre-deficient (Figs. 20 and 21), but some woven bone is present near the fracture line. In one fracture there is a small well defined mass of woven bone in the medullary cavity on the edge of the band of repair tissue.

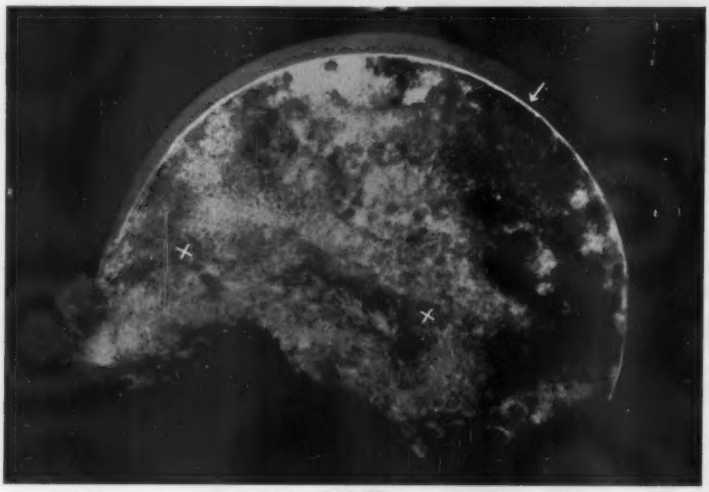

FIG. 22

Radiograph of a slice of the femur head (Case 1). This shows a normal articular cartilage with the calcified zone at its base (arrow). The normal pattern of the cancellous bone is replaced by irregularly distributed masses of calcified tissue (collections of partially calcified trabeculae in sections). Towards the base of the head is a rarefied tract (X, X) corresponding to a band of fibrous tissue. (×2·8.)

Femur head in Case 1—This shows a severe grade of the defect, and the picture is further complicated by a fracture through the base of the head which has united, mainly by fibrous tissue (Fig. 23).

The articular cartilage is intact, and shows no abnormality; with polarised light it shows the normal sheen produced by the fibrillary structure. Reticulin methods (Gömöri's and Laidlaw's) further confirm the normality of its fibrillar structure. It shows the normal calcified zone at its base in the radiograph (Fig. 22).

Virtually the whole of the cancellous bone is abnormal, being fibre-deficient and imperfectly calcified; only a few scattered fragments of the original lamellar bone remain, being incorporated in trabeculae of abnormal bone or attached to the cartilage at the chondro-osseous junction. The only other bone with a normal fibre structure is in relation to the irregular band of fibrous tissue which marks the line of union of the fracture; it consists of scattered groups of woven bone trabeculae lying in or at the edge of the fibrous tissue. A few small foci of fibrocartilage are also present in this tissue.

The trabeculae of abnormal bone are much thinner than those of the lateral condyle described earlier, their thickness rarely exceeding the normal range, and although they show abnormally wide osteoid margins these do not attain the great width of those seen in the lateral condyle. The bone tissue is very irregularly distributed, forming closely packed masses of trabeculae in some regions, while in others there are areas of porosis containing only sparsely scattered very fine trabeculae (Figs. 22 and 23).

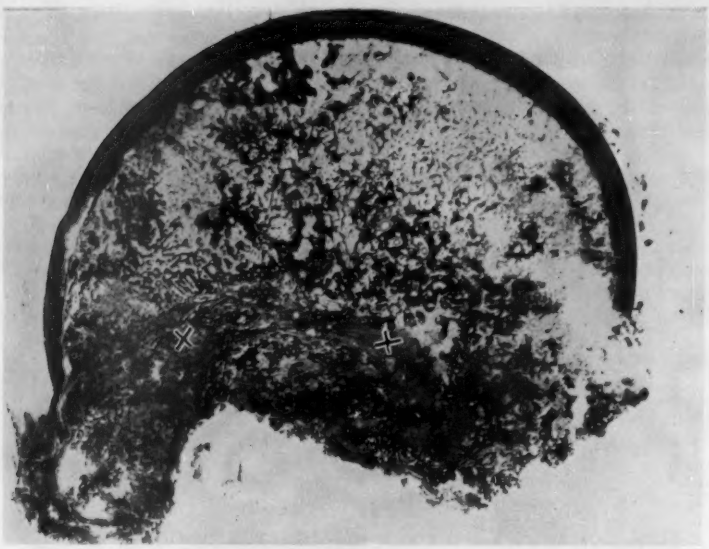

FIG. 23

Section of the same slice showing very irregularly distributed cancellous bone composed in some places of compact masses of trabeculae, but in others of more widely spaced fine trabeculae. There is a band of fibrous tissue (X) uniting a fracture through the base of the femur head. (Both calcified and uncalcified bone show up black in this figure.) (Paraffin section; Gömöri's reticulin method, ×2·5.)

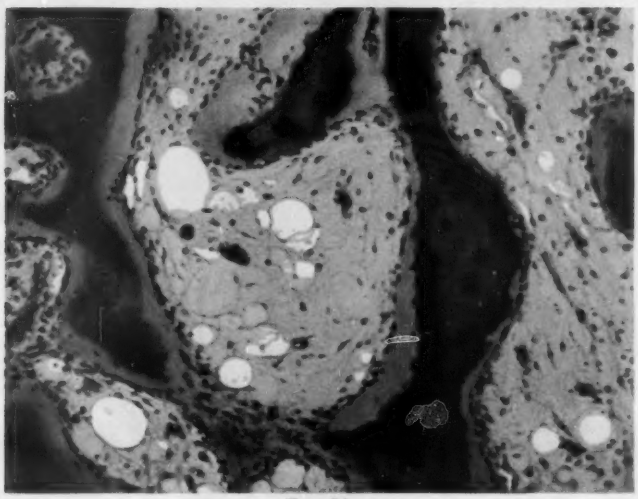

FIG. 24

Femur head (Case 1) showing trabeculae with calcified cores and wide osteoid borders. The calcified core of the trabecula in the right half of the field shows very dark blue margins: this is recent granular calcification at the edge of an older calcified fragment which shows pitted margins where these are not obscured by the recent calcification. To the left are some recently formed trabeculae with incomplete calcification of their centres where osteocytes are numerous and crowded together as in woven bone (see text). All the bone and osteoid seen here is fibre-deficient. The cancellous spaces are filled by gelatinous marrow containing many fibroblasts: this has replaced most of the fatty marrow, only a few groups of fat cells remaining.

The largest of these areas of porosis (right of Figs. 22 and 23) corresponds with the non-weight-bearing region of the femur head.

The cancellous spaces contain fatty marrow, but much of this has been replaced by gelatinous

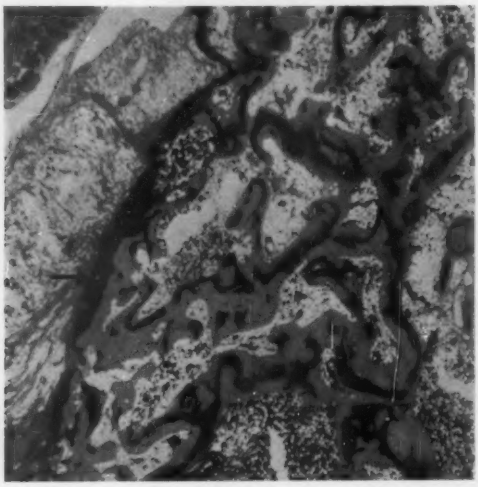

FIG. 25

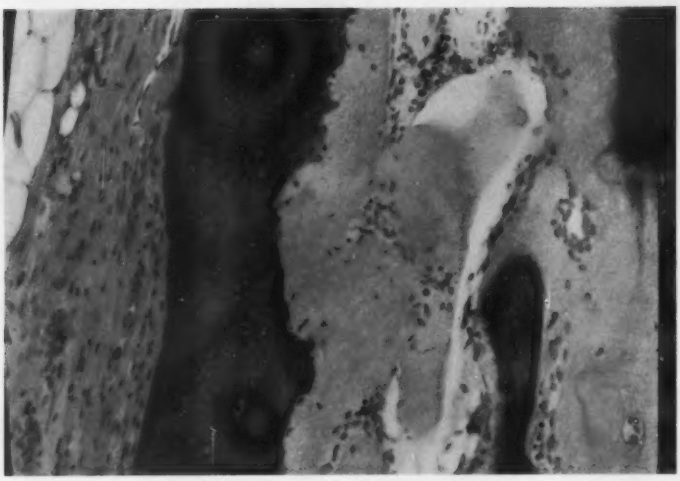

FIG. 26

Greater trochanter of femur (*Case* 2) *showing outer shell of metaplastic bone.* Figure 25 shows the outer thin shell of metaplastic bone (arrow) described in the text. Inside this (to the right) are irregular trabeculae of very poorly calcified (osteoid tissue containing dark strands of calcification) fibre-deficient bone. (Celloidin section: Ehrlich's haematoxylin and eosin, ×11.) Figure 26 is part of Figure 25 showing periosteum on the left with the shell of calcified (dark) metaplastic bone attached to it. The inner (right hand) surface of the bone is very irregular, being pitted by numerous Howship's lacunae. Attached to this surface is a layer of pale-staining fibre-deficient osteoid tissue. To the right of the figure is some more similar tissue with two deeply stained calcified areas. (Section as Figure 25, ×160.)

marrow in which are numerous fibroblasts with, however, few fibres except in the region of the fracture site. Scattered mononuclear cells are also present, and in some areas collections of macrophages containing blood pigment. Figure 24 shows the appearance of some of the trabeculae of abnormal

bone with wide osteoid margins, and also that of the gelatinous marrow with fibroblasts, which has replaced most of the fat cells in this area.

Small scattered foci of active osteoclasis are present on the bone surfaces throughout the section. *Comment*—The repair tissue in the region of the fracture is similar to normal callus; many of the areas of closely packed trabeculae of abnormal bone have probably replaced earlier abnormal bone and are also secondary to injury, as is the fibroblastic reaction in the marrow spaces. Reactions to crumpling and fractures are discussed more fully later.

Femur head in Case 2—The piece sectioned was about half the femur head with its articular cartilage.

The appearances are essentially similar to those seen in the femur head of Case 1. There is no fracture, but there is evidence of reaction to damage.

The articular cartilage shows no abnormality. With the exception of a few scattered fragments the original lamellar bone has disappeared, being replaced by trabeculae of fibre-deficient bone with wide osteoid margins. Much of the fatty marrow has been replaced by gelatinous tissue containing many fibroblasts with scattered mononuclears, many of which are macrophages containing blood pigment.

FIG. 27

Figure 27 shows the same field viewed by polarised light. The periosteum, on the left, is seen to be made up of coarse birefringent fibres which bend over towards the right as they approach the line of dark staining calcification which marks the periosteal boundary of the bone. This deeply stained line obscures the brightness of the fibres but they continue without interruption into the metaplastic bone. The fibres are cut off abruptly at the inner (right) irregular surface of the bone. The bone tissue in the right half of the figure being all fibre-deficient shows no birefringence.

At one side of the section is an area (approximately $1{\cdot}0 \times 1{\cdot}5$ centimetres) containing many collections of macrophages filled with blood pigment; there are some small foci of fibrosis. Many groups of woven-bone trabeculae are also found: when viewed with polarised light these show a normal or nearly normal fibre content. Where the fibre content is good the bone appears well calcified without excessively wide osteoid margins.

Small scattered foci of active osteoclasis are found on the bone surfaces throughout the section. *Comment*—The femur head is very similar to that in Case 1, showing a severe grade of the defect, and also evidence of damage, but without obvious fracture. The damage is doubtless produced by normal stresses on the soft and easily compressible bone. The formation of woven bone in the area showing evidence of damage is in keeping with the findings at other sites.

Greater trochanter in Case 2—Sections of the greater trochanter, which was compressible by the fingers and easily cut with a knife, show a very thin (0·02–0·2 millimetre) shell of calcified metaplastic bone containing numerous fibres continuous with those of attached ligaments and fasciae (Figs. 25 to 27). The inner surface of the shell appears as a sharply defined irregular line pitted by numerous Howship's lacunae, osteoclasts being still present at a few points. Abnormal bone has been deposited

on this surface, and replaces the whole of the normal cancellous bone. The area examined covers a complete cross-section of the trochanter, and over rather less than half this the cancellous bone is in the form of thick irregular trabeculae largely composed of osteoid tissue, as in the lateral condyle (Figs. 10 and 25). Fatty marrow, with a little haemopoiesis in places, fills the cancellous spaces. In the rest of the cross-section the trabeculae are similar, but are smaller and more closely packed, greatly reducing the size of the cancellous spaces. There is also evidence of trauma (produced no doubt by distortion of the soft bone) as shown by recent haemorrhages, collections of macrophages containing blood pigment, proliferation of fibroblasts and, in places, a little fine fibrosis. These changes are largely confined to the cancellous spaces in the area of closely packed trabeculae. The thin shell of metaplastic bone over this area is much distorted.

Comment—The trochanter shows similar changes to those in the lateral condyle, together with traumatic effects.

Vertebral body (L.5) in Case 2—The section covers rather more than half the vertebral body with most of an attached intervertebral disc. Virtually the whole of the bone, with the exception of a few fragments forming an incomplete paper-thin layer on the surface, is of the abnormal type. This forms, in the central region of the vertebral body, irregular, only partly calcified, coarse trabeculae with wide intervening marrow spaces, similar to those seen in the femur condyle (Fig. 24). Abutting on the intervertebral disc is a transverse band approximately 0·5 centimetre in vertical depth, composed of more closely packed irregular masses of the partially calcified trabeculae. This zone of more compact tissue is responsible for the band of increased density seen on each side of the intervertebral discs in the clinical radiographs of this case. The marrow spaces contain haemopoietic marrow with diffusely scattered fat cells.

LESS ADVANCED GRADE OF THE DEFECT

The specimens from rib and skull of Case 1 show less severe changes than any of the other samples examined.

Rib in Case 1—The rib (described earlier) appeared normal externally, with a firm cortex, but showed numerous chalky nodules in the medulla (Fig. 28). Figure 29 gives a general view of a transverse section and shows the calcified nodules which appear as irregular darkly staining masses springing from the cortex of the outer side of the rib and encroaching on the medullary cavity. These masses are composed of fibre-deficient bone matrix, here showing much coarsely granular deeply haematoxyphil calcification. Trabeculae of the cancellous (lamellar) bone are seen at the upper and lower poles of the medullary cavity where they are partly surrounded by the abnormal bone; elsewhere they have been almost completely absorbed. Haemopoietic marrow, partly fatty, occupies the available medullary space. The cortex as a whole is intact and not porotic, but focal absorption of bone has occurred at many spots. This focal absorption is of interest because it illustrates the early stage of the process by which the normal lamellar bone of the skeleton is replaced by the abnormal bone tissue. A group of small cavities—" Haversian spaces "—produced by osteoclastic absorption starting on the walls of Haversian canals—is well seen in the region of the cortex marked X in Figure 29. Figure 30 shows part of this region more highly magnified. It shows well the rounded spaces with dark margins crenated by innumerable Howship's lacunae. The spaces are lined wholly or in part by a layer of lightly staining fibre-deficient osteoid tissue which has been deposited on the eroded (lacunar) surfaces of the lamellar bone. In the cavities is some reticular marrow tissue containing small vessels. (This has pulled away from the wall of the cavity owing to shrinkage of the section, carrying with it the thin line of endothelial-like osteoblasts which covered the osteoid tissue.) As the fibre-deficient osteoid tissue matures, it calcifies to a variable extent, showing deeply haematoxyphil granular calcification which usually starts at the junction of the osteoid tissue with the lamellar bone (as seen on the upper

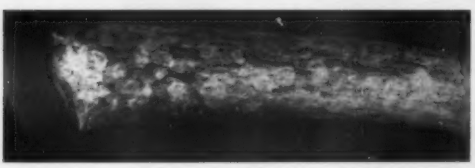

FIG. 28

Radiograph of rib from Case 1 showing the calcified nodules seen in the medullary cavity of the specimen.

FIG. 29

Transverse section of rib (Case 1). The cortex, which is of about normal thickness, is composed of lamellar bone, which has, however, undergone some local absorption with the production of rounded spaces: (a group of these is seen opposite X). The calcified nodules seen in the radiograph appear here as nodular masses attached to the medullary surface of the lateral cortex (right of Figure) and protruding into the medulla. They are composed of fibre-deficient bone matrix showing irregular areas of deeply haematoxyphil coarsely granular calcification. (Paraffin section; iron haematoxylin and eosin, ×16.)

and left-hand wall of the large oval space nearest the top of the figure). As it increases in amount, the abnormal tissue may fill up the Haversian space, always, however, leaving one or more vascular canals. Such a filled-up space showing three vascular canals lying in fibre-deficient bone formed by calcification of the osteoid tissue is seen in the top left corner of the figure. The crenated (lacunar) line forming the wall of the original space is clearly seen. The abnormal bone in this space has been eroded in part by the subsequent formation of the large oval cavity on its right; this cavity has also encroached on the small Haversian system farther to the right.

Skull in Case 1—The specimen from the skull vault, though it showed no naked-eye or radiographic abnormalities, shows deposits of fibre-deficient osteoid (slightly calcified in

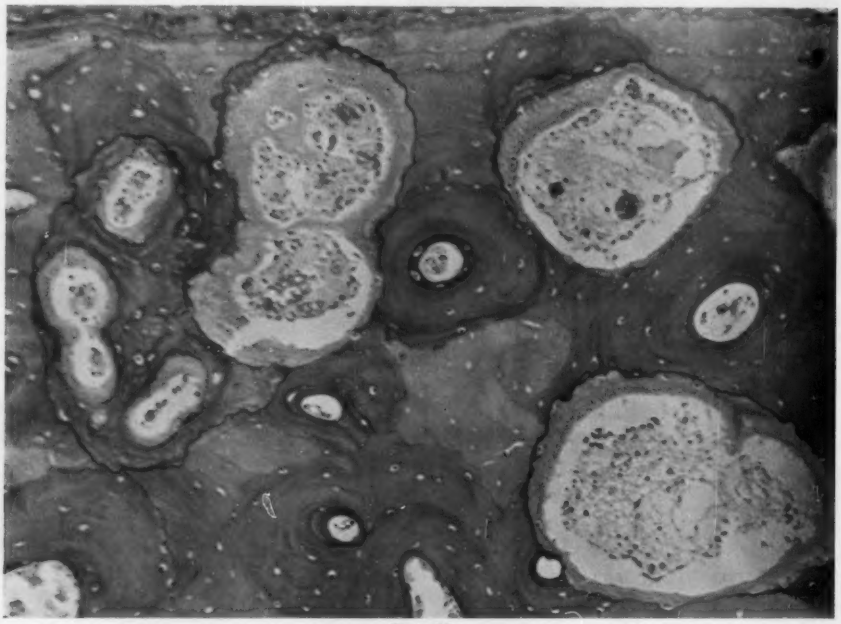

FIG. 30

To show the early stage of the replacement of lamellar bone by fibre-deficient bone. A more highly magnified view of the area opposite X in Figure 29. The lamellar bone of the rib has been hollowed out by osteoclasis to form the three larger cavities seen in the figure. The walls of these cavities show deeply stained crenated outlines produced by numerous Howship's lacunae. On the inside of the irregular surface a layer of pale staining fibre-deficient osteoid tissue has been deposited; this shows in the left-hand space some dark staining calcification in the older part next to the wall of the space. To the left of the figure is a fourth cavity largely filled in by calcified fibre-deficient bone but leaving three smaller spaces within it lined by fibre-deficient osteoid tissue. (Section as Figure 29, ×170.)

places) on the surfaces of the trabeculae. The deposit forms layers of different thicknesses on the walls of the marrow spaces, which in places it completely fills, apart from one or more small vessels, replacing the haemopoietic marrow. The deposit is distributed in a patchy manner and with a hand lens is seen in haematoxylin and eosin sections (Fig. 31) as islands of pink tissue contrasting with the deeply stained haemopoietic marrow which fills the surrounding cancellous spaces. There is very little disturbance of the architecture of the bone, which shows no appreciable porosis. With a higher magnification (Fig. 32), however, it can be seen that lacunar absorption has taken place on many of the bone surfaces now covered by the abnormal tissue; in other places the tissue has been deposited on the smooth uneroded bone surface.

In addition to the more obvious changes described above, partial defects were discovered in the Haversian systems of both the skull and the rib of Case 1; these are described later.

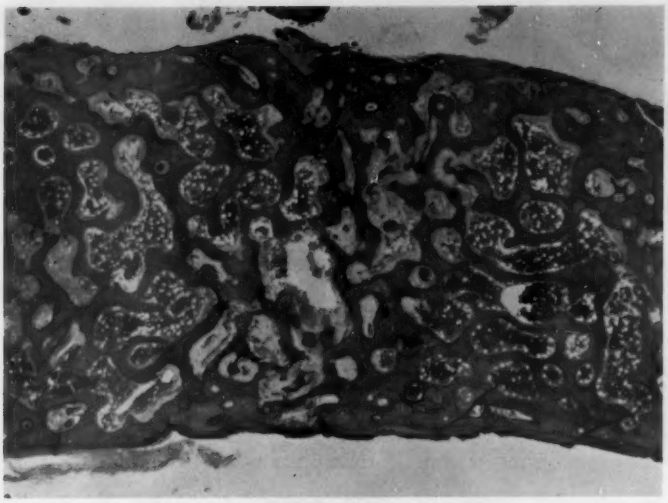

FIG. 31

Section of the skull (Case 1) which showed the earliest stage of the defect found in the material examined. There is no appreciable disturbance of the architecture of the bone (the specimen showed no abnormality on naked-eye examination or radiographically). The cancellous spaces on both sides of the figure are filled by a mixture of fatty and haemopoietic marrow but those in the centre are filled by a pale-staining tissue, found to be fibre-deficient osteoid tissue showing calcification in places. (Celloidin section; Ehrlich's haematoxylin and eosin, ×11.)

FIG. 32

Part of the same section as shown in Figure 31. It shows fibre-deficient osteoid tissue, with some calcification (dark patches), filling a small cancellous space. There has been no appreciable erosion of the wall of this space but the neighbouring one, also filled by abnormal osteoid tissue, shows some Howship's lacunae in the region marked by an arrow. (Section as in Figure 31, ×200.)

MODE OF REPLACEMENT OF THE NORMAL BY ABNORMAL BONE

In all the sections examined there is evidence of removal of the lamellar bone by lacunar absorption (removal by osteoclasts with the formation of Howship's lacunae). The process is active in some areas with numerous osteoclasts in the sections; in others there is evidence of past osteoclastic activity with Howship's lacunae no longer containing osteoclasts. In some regions there is hardly any lamellar bone, but the few remaining fragments show clear-cut lacunar margins. The early stage of the process is well shown in Figure 30, a transverse section of rib cortex from Case 1 just described.

In addition to removal of lamellar bone there is a considerable amount of osteoclastic removal of the new abnormal bone, both calcified and non-calcified. This is very active in the lateral condyle of femur in Case 2 (Figs. 11 and 12), where thick trabeculae of partly calcified abnormal bone are hollowed out in many places; the hollows are filled by a cellular connective tissue with numerous osteoclasts at bone surfaces.

Hollowing out of the trabeculae in this way—so-called "dissecting absorption"—is found in hyperparathyroidism and also in some cases of osteomalacia (Wernly 1952). In this material it is obvious only in the lateral condyle of the femur and in a few small foci in the vertebral body (Case 2). There is no other evidence suggesting osteitis fibrosa in spite of the advanced state of the bone changes. Primary hyperparathyroidism can therefore be excluded. At most there could be a mild grade of secondary hyperparathyroidism but there is scanty evidence for this.

FINER HISTOLOGICAL STRUCTURE OF THE DEFECTIVE BONE

In addition to the absence (or virtual absence) of birefringence with polarised light and the absence of fibre structure with Schmorl's Thionin method, the abnormal fibre-deficient tissue shows a diminished affinity for most of the usual histological stains. It stains poorly with eosin and similar acid stains and also with the standard connective tissue stains such as van Gieson's, Mallory's, Heidenhain's azo-carmine and Masson's trichrome. Of these connective tissue stains Heidenhain's azo-carmine and a modification of the Masson-Golden method, using aniline blue instead of green as the collagen stain, show the structure best but none shows the fine detail which can be obtained by the silver methods for reticulin.

The following description is based mainly on the study of thin paraffin sections from Case 1 stained by Gömöri's reticulin method. In satisfactory sections stained by this method normal bone is aureophil and the fibre structure of both lamellar and non-lamellar (woven) bone shows up well. The marked difference between the abnormal bone matrix and normal lamellar bone is well demonstrated in Figures 33 and 34 from a Gömöri-stained section showing islands of normal lamellar bone in a trabecula of fibre-deficient bone matrix (femur head Case 1).

In Figure 33 two fragments of normal bone show up as compact aureophil tissue with lamellar markings; these fragments lie in fibre-deficient bone matrix consisting of a much less compact tissue showing a feltwork of fine fibrils on which is superimposed a net of rather thicker fibres, some aureophil, many argyrophil. The cell spaces (the contained osteocytes being, of course, unstained by this method) appear as hollow capsules connected to the fibre net. It can be seen that an irregular area showing a prominent argyrophil net surrounds the two fragments of normal bone and extends as a broad band between them. In the upper part of the section are two areas (one to the left and the other to the right of the smaller bone fragment) where the fibre net is aureophil and therefore stands out much less prominently than the argyrophil net. The areas showing the prominent argyrophil net are those in which granular strongly haematoxyphil calcification is present in a contiguous serial section stained with haemalum and eosin. The areas that are predominantly aureophil are those showing the uncalcified fibre-deficient osteoid tissue. This difference in the staining reaction of fibrils and fibres in calcified and non-calcified areas is apparent in all the material studied by reticulin methods (see also Figures 34 and 42).

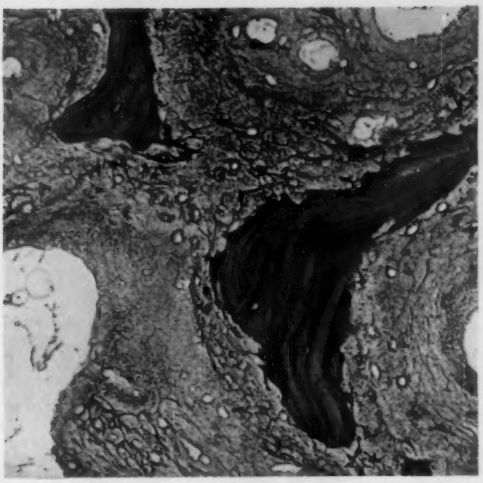

FIG. 33

Showing the finer structure of the fibre-deficient bone. Part of a trabecula from the femur head of Case 1 showing two fragments of normal lamellar bone surrounded by fibre-deficient bone. The normal bone shows a compact fibre structure with lamellar markings and is red (aureophil). The surrounding abnormal bone shows a meshwork of fine fibrils many of which are black (argyrophil) particularly in a zone round the two fragments of bone and in the area between them. (Paraffin section (5μ); stained by Gömöri's method, $\times$170.)

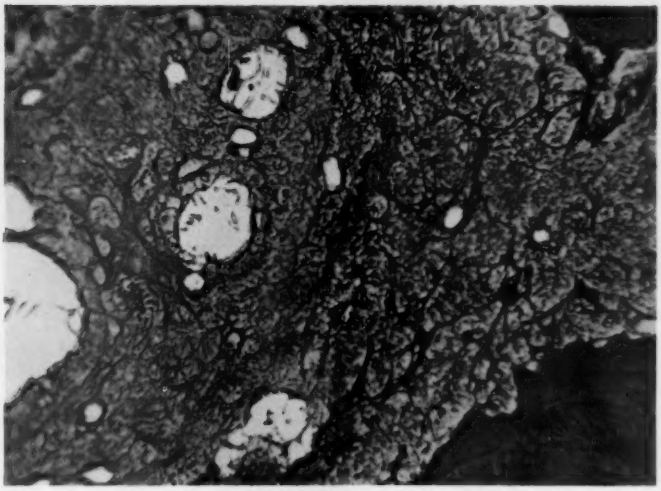

FIG. 34

Part of Figure 33 more highly magnified. In the right half of the figure is part of the argyrophil area extending between the two fragments of lamellar bone (seen in right-hand corner). It shows prominent black fibres on a finer fibrillary background. To the left is an aureophil area showing rather less clear-cut aureophil fibres with a fibrillary background. In both areas the walls of the rounded cell spaces are usually connected to the fibre net. (The same section as Figure 33, $\times$500.)

Figure 34, a higher magnification of part of Figure 33, shows an aureophil area (top left in Figure 34) and part of the argyrophil area which extends round the two lamellar bone fragments, parts of which are seen in the right-hand corner of this figure. Both these areas show a fibrillary background and a superimposed net of fibres with scattered cell capsules attached to it. In the aureophil area the net is less prominent, being more lightly stained and less sharply defined than in the argyrophil area; some of the cell capsules in the aureophil area are partly argyrophil.

The above example holds good for all the abnormal bone matrix in these cases but the amount of collagen fibre varies from place to place according to the local severity of the defect; where the defect is maximal only a tenuous fibrillary feltwork is present. Well defined fibres become more plentiful where the defect is less complete but the structure still remains very deficient in collagen fibres and far removed from that of either lamellar or woven bone except in a few areas where there is a reaction to injury.

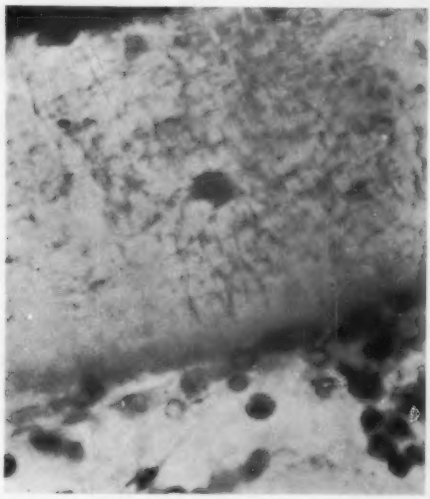

FIG. 35

FIG. 36

Figure 35—Showing an osteocyte with faintly stained canaliculi (or cell processes?) in fibre-deficient osteoid tissue (greater trochanter, Case 2). (Celloidin section; Schmorl's Thionin, ×620.) Figure 36—Osteocyte in an area of calcified fibre-deficient bone (rib, Case 1) showing the cell space as a clear area with irregular channels (canaliculi) emerging from it. (Paraffin section; iron haematoxylin and eosin, ×700.)

Argyrophil staining of collagen fibres and fibrils in areas of haematoxyphil granular calcification—The argyrophil reaction of collagen fibres and fibrils in areas of haematoxyphil granular calcification as compared with their aureophil staining elsewhere has, so far as I know, not been previously noted. It was difficult to be sure that this marked difference in the appearance of such calcified areas did not correspond to a real difference in fibre content; but it was found that the change from an aureophil to an argyrophil reaction can be seen in a single fibre as it passes from the non-calcified to the calcified region. This change is seen not only in the abnormal bone matrix but in normal collagen bundles of ligaments and fasciae which change from aureophil to argyrophil as they enter the calcified zone which is commonly present close to their bony attachments.

OSTEOCYTES AND OSTEOBLASTS IN THE ABNORMAL BONE

Defective staining of canaliculi—The most obvious abnormality in relation to the bone cells is the failure (complete or partial) of Schmorl's Thionin method to stain the canaliculi in the fibre-deficient bone and osteoid tissue.

In normal bone Schmorl's Thionin appears to stain the wall of the canaliculus, and the stain also fills the lumen. The cell processes are also stained (a lighter blue) and can be seen in some cells

as they leave the cell to enter the mouth of the canaliculus, but cannot be seen in the canaliculus, the lumen of which is blotted out by dense staining (apparently a precipitate of the stain).

In the abnormal bone Schmorl's stain usually shows either no canaliculi or very faintly

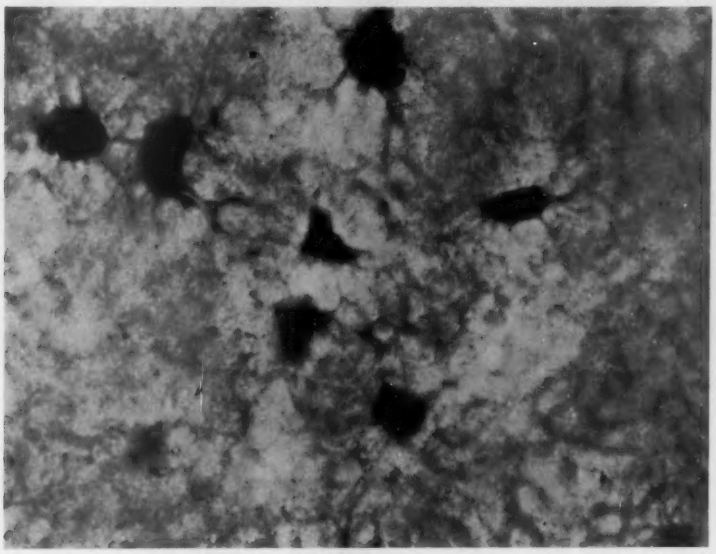

FIG. 37

Osteocytes with cell processes in fibre-deficient osteoid tissue. In the background of the figure is a faintly stained network (abnormal canaliculi?). From the lateral condyle of femur (Case 2). (Celloidin section; Schmorl's Thionin, ×1,250.)

FIG. 38

A group of osteocytes showing capsules of cell spaces with emerging rays forming the "asterisks" described in the text. These are in fibre-deficient osteoid tissue (femur head, Case 1). (Paraffin section; Gömöri's reticulin method, ×600.)

stained ones, which are visible only with fairly high magnification (Fig. 35). The staining is usually patchy, so that the canaliculus appears as a broken line. It looks as though the wall did not stain but as if a scanty deposit of stain formed in the lumen (possibly on the surface of the cell process). There is no doubt that cell processes are present in the abnormal bone,

though the channels in which they run do not appear to be normal canaliculi. Further information on this point is obtained with reticulin stains.

Relation of the osteocytes to the fibre structure of the defective matrix—As already noted, the bone cells usually lie on the strands of the fibre net where this is present, the fibres also forming " capsules " round the cell spaces (Figs. 33 and 34). In areas where the defect is severe Gömöri sections often show these capsules with rays passing out from them—like minute stars or " asterisks " (Fig. 38). The rays of the " asterisks " are hollow, as demonstrated in Figure 39, a canaliculus passing down the centre of each to enter the cell space. There is in normal bone a specialised very thin ensheathing layer lining the cell spaces and forming the

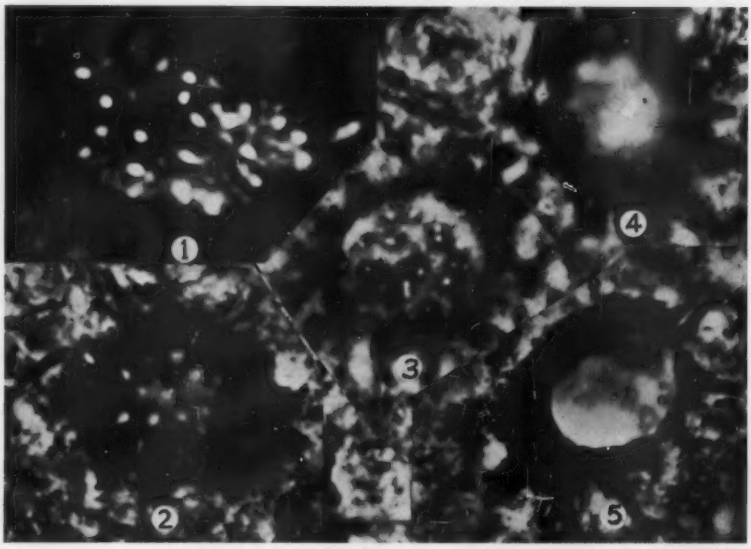

FIG. 39

Demonstrating that the " rays " of the " asterisks " seen in Figure 38 *are pierced by canaliculi.* Composite picture of four cells in fibre-deficient osteoid tissue and one in normal lamellar bone. (1) Osteocyte in normal lamellar bone, section passing tangentially to the wall of the cell space and showing openings of emerging canaliculi. No details are visible in the surrounding dense collagen of the bone matrix. (2, 3 and 4) Fibre-deficient osteoid tissue showing cell spaces with their surrounding walls and emerging "rays" cut in different places. (2) and (3) show emerging canaliculi cut transversely; in (3) the walls of the canaliculi which form the " rays " and the " asterisks " are well seen. The surrounding bone matrix shows very scanty fibres as compared with (1). (5) A similar cell space with a well defined wall and emerging rays. One of these (top right) is cut longitudinally showing the central canaliculus emerging from the cell space. (Paraffin sections (5μ): Gömöri's reticulin method, $\times$2,300.)

walls of the canaliculi. The " asterisks " are evidently formed of this ensheathing layer, which here stands out prominently owing to the dearth of collagen fibre in the surrounding matrix. The layer is not, however, continued along the whole course of the canaliculus, as in normal bone, but tails off and becomes incomplete at a short distance from the cell, appearing only as argyrophil fibrils connecting neighbouring cell spaces (Fig. 38). The " asterisks " stain like collagen—red with van Gieson's stain and blue with Heidenhain's Azan or similar stains containing aniline blue—but with these stains they do not present the clear-cut outlines seen by Gömöri's method.

These findings suggest that the bone cell determines the formation of the wall of the cell space and canaliculus but here, owing to some defect in the mechanism of bone collagen

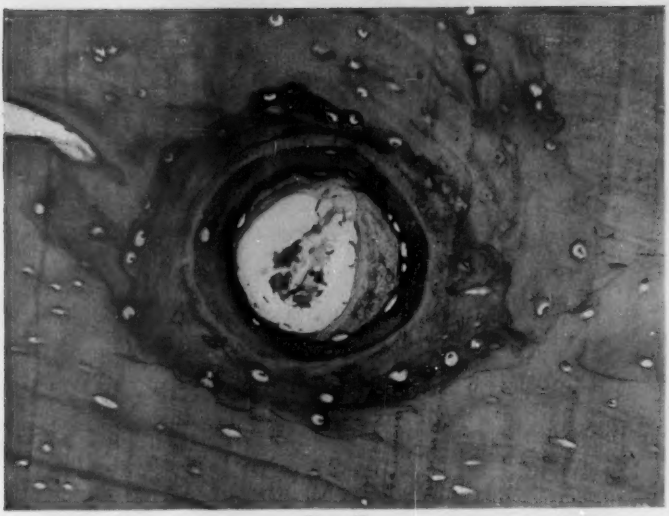

FIG. 40

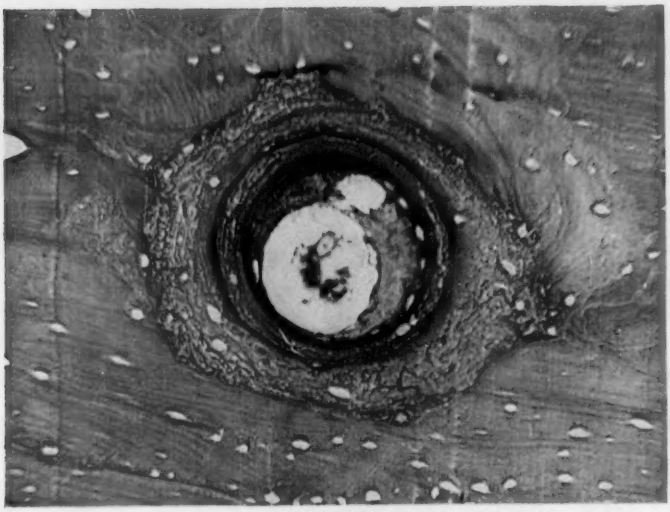

FIG. 41

Showing a partially defective Haversian system in the skull of Case 1. Figure 40 shows a Haversian system most of which is haematoxyphil, staining blue in contrast to the pink (eosinophilic) staining of the surrounding normal bone. Within the blue system and concentric with it is a well marked eosinophilic band. Two less clearly marked incomplete bands are seen on each side of the upper part of the main band. (Paraffin section; haematoxylin and eosin stain, ×220.) Figure 41—A neighbouring serial section showing the same Haversian system stained by Gömöri's reticulin method. This shows that the Haversian system is largely made up of fibre-deficient bone composed of a meshwork of argyrophil fibril. But the eosinophilic ring and the partial rings on each side of it in Figure 40 are seen to be aureophilic like the surrounding bone.

production, its influence is effective only in the immediate vicinity of the cell—beyond this the canalicular walls are imperfect.

Note on the layer of bone matrix forming the walls of the cell spaces and canaliculi—The cell spaces and canaliculi in bone are covered by a special boundary layer which stains differently from the bulk of the bone matrix. Recently deposited immature bone matrix (osteoid) stains like collagen fibres (red with van Gieson's stain and blue with Heidenhain's Azan or similar connective tissue stains containing aniline blue) but more mature bone does not; it stains yellowish (picric acid) with van Gieson's stain and red with Heidenhain's Azan. The very thin layer of matrix surrounding the cell spaces and forming the walls of the canaliculi reacts to connective tissue stains like recently formed osteoid tissue and never " matures " like the rest of the bone matrix. This layer can be stained black by silver impregnation (Weidenreich 1930) and often shows up black in Gömöri-stained sections. The rays of the argyrophil " asterisks " described above are clearly composed of this layer.

Distribution and form of the osteocytes—The absence of fibre systems in the defective bone removes the main basis for distinguishing between lamellar and woven bone. The form and distribution of the osteocytes is, however, different in these two types of bone; they are more uniform in size and distribution and fewer in number in lamellar bone as compared with woven bone. On the basis of its cytology most of the abnormal bone is defective lamellar bone; the osteocytes being, for the most part, sparsely and fairly uniformly scattered as in lamellar bone. (Cell counts of lamellar bone and fibre-deficient bone in a section of the femur shaft of Case 2 showed virtually the same number of osteocytes per unit area.) This conclusion is further supported by the fact that even where the defect is severe, as in the lateral condyle of the femur in Case 2, a trace of lamellar structure is revealed in a few places in the abnormal matrix by the use of polarised light. (See also the description of partial defects in lamellar systems, which follows.)

Abnormal bone with osteocytes crowded together like those of woven bone is found at sites where there is evidence of trauma, and gradations in structure between this and normal woven bone may be found. Trabeculae with incompletely calcified cores with crowded osteocytes of this type are found in the femoral heads, while the osteoid tissue deposited on the surfaces of the cores shows the cellular character of lamellar bone. This corresponds with the normal sequence where lamellar bone forms on the surface of woven-bone trabeculae.

Osteoblasts—The cells on the surface of the fibre-deficient osteoid tissue form, in many places, a single layer of flattened endothelial-like cells like those on the surface of inactive normal bone (without osteoid borders). In other places the cells are plumper and may form a layer several cells thick, as seen on the osteoid borders or normal growing bone. There is therefore no obvious morphological abnormality in the osteoblasts.

Such abnormalities as have been noted in the osteocytes appear to be the result rather than the cause of the abnormal fibre structure.

PARTIAL DEFECTS IN THE HAVERSIAN SYSTEMS OF THE RIB AND SKULL (CASE 1)

The appearances to be described here were first noticed in paraffin sections of formol-fixed material from the rib and skull in Case 1 which had been decalcified in nitric acid to the point where the normal lamellar bone was eosinophilic while the granular calcification in the abnormal bone matrix deposited in the cancellous spaces remained strongly haematoxyphil. In these sections it was noticed that some of the Haversian systems in the apparently normal cortical bone remained haematoxyphil in contrast to the surrounding lightly eosinophilic systems. Some of these " blue systems " showed also interruptions in their concentric pattern. On further investigation of this appearance in serial sections stained by haemalum and eosin, by Gömöri's reticulin method, by van Gieson's stain, and by Heidenhain's azo-carmine, it was found that in many of the Haversian systems there were interruptions in the regular concentric arrangement of the lamellar bone due to local failure of the normal collagen

production. Figure 40 shows the increased haematoxyphil staining of such a Haversian system as compared with the surrounding bone; within the blue system, however, are some concentric groups of eosinophilic lamellae. The Gömöri-stained section of this system (Fig. 41) shows that much of the bone is abnormal, presenting a dark granular-looking appearance; but the concentric groups of lamellae which are eosinophilic in Figure 40 show the normal aureophil collagen staining.

A similar defect is seen in more detail in Figure 42, which shows part of a Haversian system, including part of the central canal. The aureophil area to the right shows the normal appearance of lamellar bone; between this and the central canal are two concentric zones

FIG. 42

A more highly magnified view of the inner part of a partially defective Haversian system similar to that seen in Figure 41. The right-hand side of the figure shows normal lamellar bone. There are then two zones of defective bone alternating with rings of normal bone. The inner ring is the wall of the central canal on which a relatively thick layer of abnormal bone has been deposited (left of figure). (Paraffin section; Gömöri's reticulin method, ×700.)

of defective bone showing networks of argyrophil fibrils separated by a band of normal aureophil bone. Another band of normal bone forms the wall of the central canal, within which is a more recent deposit of abnormal bone matrix, the older part of which (nearest the wall of the canal) shows a net of clear-cut argyrophil fibrils while the most recently deposited part (bottom left in Figure 42) shows a lightly aureophil meshwork of less sharply defined fibrils. This aureophil network is uncalcified, but all the areas with argyrophil fibrils show granular calcification in haematoxylin and eosin sections.

There is no sharp break, by a cement line, between the rings of normal bone and the

defective bone in these Haversian systems—the normal bone has not therefore been eroded and replaced by the abnormal as in most places. Here the process of deposition has been uninterrupted; but the quality of the bone laid down has varied from time to time. (This is

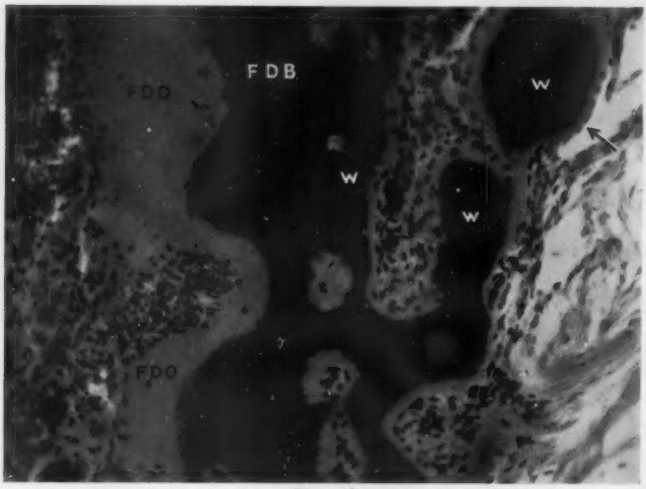

FIG. 43

FIG. 44

Fracture callus (Case 2) showing woven bone with good fibre content and normal osteoid borders while the adjacent fibre-deficient bone has wide osteoid borders. Figure 43—To the right, well calcified woven bone (W) with normal osteoid borders (well seen opposite arrow). To the left fibre-deficient bone (FDB) with wide osteoid borders. (Celloidin section; Ehrlich's haematoxylin and eosin.) Figure 44—The same field using the polariser. The narrow osteoid borders of the woven bone show birefringence (well seen opposite arrow) but the wide border of fibre-deficient osteoid tissue (FDO) does not.

clear from the fact that Haversian systems result from the filling up of spaces which have been produced by osteoclastic erosion; such (Haversian) spaces are filled up by concentric rings of bone deposited in chronological order from without inwards.)

There is no evidence that these local fluctuations in the quality of the bone deposited are due to fluctuations in the disease as a whole.

This type of deposit in the Haversian systems was seen in the two least affected bone samples examined and represents the least severe (and presumably earliest) lesion observed in this material.

REACTIONS TO INJURY

Five healing fractures and also microscopic evidence of damage judged to be due to deformation of the poorly calcified bone tissue have been observed in this material. The chief point of interest in these injuries is the fact that the repair reaction has produced woven bone containing abundant collagen fibres. This is most abundant in the periosteal reaction over the rib fractures (Case 2). It is also present in smaller amounts in the deeper part of the rib callus (the bulk of which is fibre-deficient), in relation to the band of fibrous union at the base of the femur head (Case 1), and in the neighbourhood of small areas of fibrosis and blood pigment in the femur head of Case 2.

It is clear therefore that although the bone collagen is in general grossly defective, the healing process can stimulate its formation locally. (Much the same thing happens in osteogenesis imperfecta where, in spite of the generally poor production of bone, fracture callus forms without difficulty.)

A further important point is the fact that this woven bone with good collagen content calcifies normally, or nearly so, having relatively narrow osteoid margins in contrast to the very wide margins seen in the adjacent fibre-deficient bone (Figs. 43 and 44).

This shows that the failure of calcification is dependent on the defect in the bone matrix, and is not a general humoral one as in osteomalacia. (In fractures of osteomalacic bones the condition is of course the reverse of that seen here, the newly formed woven bone showing less calcification than the surrounding older bone.)

Woven bone in fracture callus is normally replaced by lamellar bone as healing advances. The process starts quite early by the deposition of lamellae on the surfaces of woven-bone trabeculae which have often been partly eroded (surfaces pitted by Howship's lacunae). The same process occurs in these rib fractures but instead of lamellar bone fibre-deficient bone is deposited on the woven-bone surfaces.

HISTOLOGY OF SOFT TISSUES

Case 1—Pieces of the following soft tissues (formol-fixed) were received for examination: skin nodule, right lung apex, liver, spleen, kidney, adrenal, uterine fibroid, nodule from falx cerebri, basilar vessels, brain stem and pituitary. Microscopic examination confirmed the necropsy findings already outlined. Apart from the xanthomata on the skin there was no evidence of storage of lipoid or other abnormal substance. (The possibility of skeletal lipoidosis was considered clinically.)

Case 2—Sections of the following soft tissues were examined microscopically: thyroid, axillary gland, liver, kidneys, spleen, prostate, left testis, epididymis and vas, marrow from sternum and femur. No significant lesions other than those confirmatory of the gross findings already outlined were discovered.

In neither case were the parathyroids examined histologically; in Case 2 one parathyroid (not enlarged) was found but not sectioned.

When the collagen fibre defect in the bone matrix was discovered a careful examination of the connective tissue structures in all the soft tissue material was made, but revealed no abnormality. As already noted, the connective tissue of the periosteum and such attached ligaments and fasciae as are present in the sections show no collagen fibre defects.

The walls of the various blood vessels included in the sections examined, both outside and inside the bones, show no defect in the formation of the connective tissue of their walls, and there is no evidence of spontaneous haemorrhage from them.

PROTEIN CONTENT OF THE FIBRE-DEFICIENT BONE

One of the chalky nodules from the rib in Case 1 was removed and analysed for calcium and nitrogen, the nitrogen figures being a measure of the protein content.

The method used was that described by Baker, Butterworth and Langley (1946) and the result was compared with the figures for the series of normal bones reported in the above article.

The calcium/nitrogen ratio for cancellous bone of normal adult rib was 5·1:1; the Ca/N ratio for the chalky nodule was 28:1. This could mean either a very densely packed mass of calcium salts (nearly five times as dense as the cortex of the normal femur) or a very low protein content. It would, of course, be impossible to pack the calcium salt as densely as this (nearly as dense as lead), but apart from this consideration the histological picture shows that the nodules are not completely calcified, there being borders of osteoid tissue considerably wider than those in normal growing bone. We are therefore justified in concluding that the calcium content did not exceed the normal, *i.e.*, the amount present in the same volume of normal bone, in which case the protein content of this fibre-deficient bone matrix must be less than one-fifth of that in the bone matrix of a normal adult rib. This figure gives a quantitative expression of the severe grade of collagen defect recognised histologically in these nodules.

DISCUSSION

While the essential histological lesion found in these cases of fibrogenesis imperfecta ossium is a defect in the fibres of the bone matrix, it is quite possible that there is also a defect in the ground substance between the fibres; but, owing to its amorphous nature, this is difficult to study histologically.

A rather surprising feature of the defect is that it should be limited to the bone collagen. Electron-microscopy does not reveal any essential difference between the collagen fibres of bone and those of the soft tissues, and the osteoblast is usually regarded as a not greatly modified connective tissue cell. But in the light of these two cases, we must assume that some special mechanism is involved in the formation of bone collagen which is not required in the production of soft-tissue collagen.

Bone matrix differs from soft connective tissue in that its collagen fibres are more compactly arranged and, in lamellar bone, in a more complex pattern; it has also a much lower content of mucopolysaccharide. The ground substance, about which little is known, is certainly less watery and tougher than that of connective tissue or cartilage, and may well be different chemically. But the unique property of the bone matrix is its ready calcifiability which is not dependent solely on a high phosphatase content (since there are other tissues rich in phosphatase which do not calcify) but also upon some other factor not yet identified.

It seems clear that the deficient calcification of the bone in these cases is secondary to the defect in the bone matrix, and is not related to a calcium and phosphorus imbalance of the body fluids as in osteomalacia. In addition to the metabolic findings of Dr Donald Hunter in Case 2, the results of which are not in keeping with osteomalacia, there is histological evidence. Wherever there is, in this material, a local production of new bone with normal or nearly normal fibre content—for example, woven bone in relation to the healing fractures—this calcifies normally, or nearly so, showing narrow margins of fibre-containing osteoid tissue, whereas the wide osteoid margins are always fibre-deficient. In osteomalacia, on the other hand, the defective calcification is related, not to the fibre content, which is normal, but to the amount of new bone formation, so that recently formed bone, such as fracture callus, shows more osteoid tissue than the neighbouring less active bone.

Complete or partial failure to calcify is found in the defective matrix produced by osteogenic sarcomas. Here, examination with the polarising microscope shows a dearth of collagen fibres. The few examples I have examined, using reticulin methods, also show a

dearth of fibres, but not the peculiar fibrillary structure seen in these cases of fibrogenesis imperfecta. The same type of strongly haematoxyphil coarsely granular calcification which characterises much of the calcified tissue in fibrogenesis imperfecta is found in the matrix of these sarcomas.

Recent electron-microscopical studies of calcified bone by Robinson and Watson (1952) show that the crystals of the calcium salt are closely attached to the collagen fibril and that they are arranged in a banded pattern corresponding to the main collagen striations. This close association between the bone salt crystals and the collagen fibre seems to be in keeping with the above evidence that the calcifiability of bone matrix is dependent on the proper development of its fibre structure.

Etiology—Nothing positive is known about the cause of fibrogenesis imperfecta ossium, but some possibilities can be excluded.

It is not a congenital defect, and it seems unlikely that it is an inherited abnormality, for the disease appeared after middle life, and the patients gave no history of bone trouble in their families.

Both patients were elderly (fifty-six and sixty-four years) and no fractures or other signs of skeletal disease developed until a few years before death.

The histological findings show that the skeleton was at the start of the disease composed of normal lamellar bone (much of the bone in the rib and skull in Case 1 was found to be normal, and remains of normal lamellar bone were found in most of the specimens examined). The normal bone had been removed by osteoclasis, partly or almost completely, according to the site, and replaced by fibre-deficient bone.

The evidence therefore points to an acquired condition which had probably arisen not much before the age of fifty years in Case 1, and after fifty in Case 2.

Histologically the condition is clearly not neoplastic or inflammatory. There was no appreciable anaemia or other evidence of serious toxic changes, and in neither case had there been exposure to industrial poisons. (The patient in Case 1 was a spinster with no occupation, and that in Case 2 a retired accountant.)

No endocrine disturbance is known to produce the bone changes observed here. Examination of the pituitary in Case 1, and of the thyroid in Case 2, revealed nothing noteworthy. The parathyroids were unfortunately not examined histologically in either case, but the histological changes in the bones are not those produced by either hyperparathyroidism or hypoparathyroidism. It is true that the material from both the cases shows an increased osteoclasis, but there is no appreciable osteitis fibrosa in spite of the advanced state of the bone defect in most of the specimens examined. The metabolic changes in Case 2 do not conform to either hyperparathyroidism or hypoparathyroidism.

Some kind of deficiency disease seems a possibility but the changes found do not fit in with any known deficiency affecting the bone or connective tissues and its differences from vitamin-D deficiency have already been stressed.

In vitamin-C deficiency there is a lack of collagen fibre formation, and also a weakness of the walls of the capillaries considered to be due to a defect in the intercellular cement substance. The lesions that occur are the result of these two defects, and involve the connective tissues and capillary walls throughout the body leading to 1) inadequate collagen fibre formation—for example, in healing wounds—and deficient bone production—for instance, in healing fractures; and 2) haemorrhages in the soft tissues and in relation to bone. Though bone formation is retarded, such bone as is produced shows no collagen fibre defect, and calcified normally. In fibrogenesis imperfecta there are no haemorrhagic lesions, and the collagen defect is limited to the bone matrix where it is the reverse of that in vitamin-C deficiency, since there is a quite abundant formation of grossly abnormal bone tissue.

In vitamin-A deficiency skeletal changes have been produced in animals by Mellanby (1944) and by Wolbach (1947). These changes, which are produced only in growing animals,

are the result of a disturbance of the normal balance between absorption and deposition of bone, and appear as abnormalities in bone growth and architecture. The bone tissue itself is not histologically abnormal and calcifies normally.

The cause of this peculiar bone disease is therefore quite obscure, and is likely to remain so until opportunities occur for the clinical and biochemical study of further cases. Its discovery would probably throw light on hitherto unsuspected mechanisms peculiar to the formation of the bone matrix.

SUMMARY

1. A description is given of the pathology of a generalised skeletal disease characterised by a defect in the formation of the collagen fibres of the bone matrix—" fibrogenesis imperfecta ossium."

2. Material from two cases, a woman of fifty-six and a man of sixty-four, was examined. All the samples of bone from both patients showed the same defect, which was severe in most of the specimens, and there was radiographic evidence of similar widespread bone changes in both cases.

3. The defect is clear-cut and striking histologically, provided that sections are examined with a polarising microscope, and/or by reticulin methods.

4. As a result of the defect in the bone matrix this fails to calcify, or calcifies imperfectly, showing wide osteoid borders as in severe osteomalacia. But the fibre defect separates it quite clearly from osteomalacia, in which the fibre structure of the osteoid tissue is normal. Moreover neither the biochemical findings (Case 2) nor the radiographic appearances correspond with those of osteomalacia.

5. The collagen fibre defect is confined to the bone matrix; no defect was found in the soft tissue collagen, and even the periosteum shows a normal fibre structure.

6. Both the clinical and the histological evidence indicate that the disease is not congenital, but was, in these two patients, apparently acquired during middle age. There was no family history of bone disease.

7. The cause of the condition is quite obscure. It is not inflammatory or neoplastic, nor is there histological or clinical evidence of a toxic origin. If it is a deficiency disease it is unlike any known vitamin or other chemical deficiency.

I am particularly indebted to the late Professor H. M. Turnbull, who collaborated in the earlier part of this work, and was joint author of a preliminary report (Baker and Turnbull 1950). His name would have been attached to the present paper but, since his health prevented him from taking any further part in the work, he preferred otherwise. The sections from Case 2 and some of those from Case 1 were prepared in his department by Mr John King, for many years his chief technician; many of those from Case 1 were prepared by my late chief technician Mr F. Ward.

I am indebted to Drs Fergus R. Ferguson and Donald Hunter for clinical and radiographic data; to Drs R. Whitehead and John Ball for reading and checking the typescript, and to Mrs Sandes-Yudenitch for secretarial help.

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THE OSTEOPOROSIS OF CUSHING'S SYNDROME

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In 1932 Cushing published a series of cases showing an interesting syndrome which he termed " pituitary basophilism " and which he ascribed to a pituitary adenoma of basophil cells. Cushing's patients, all young adults, showed the following clinical features: an unusual type of increase in subcutaneous fat, involving the face, neck and trunk; osteoporosis of the skeleton, most marked in the spine, resulting in kyphosis and diminution of stature; amenorrhoea in females and ultimate impotence in males; hirsutism involving the face and trunk in females; atrophic skin with purplish striae; hypertension; diabetes; and polycythaemia.

The condition, now known as Cushing's syndrome, has been the subject of many detailed clinical and endocrinological studies in recent years. It has become apparent that the syndrome can be associated with hyperplasia or tumour formation of the adrenal cortex in the absence of a pituitary adenoma (see for example Lescher and Robb-Smith 1935). In fact Albright (1943), reviewing work in this field, ascribed the various manifestations of Cushing's syndrome to hyperadrenocorticism, with or without a basophil adenoma of the pituitary. He put forward the view that the over-production of an adrenal hormone is responsible for inhibition of protein synthesis throughout the body, and contrasted the metabolic position with that in the adreno-genital syndrome, where different adrenal hormones are concerned, including one normally responsible for the development of pubic and axillary hair.

Cushing's original account noted that bone changes were present in six out of eight cases coming to autopsy. While the clinical and radiological aspects of the osteoporosis of the spine are now well known, there is little published information on the structural pathology of this lesion or of other skeletal changes in Cushing's syndrome. The present paper, therefore, describes the histological changes in bone in autopsy material from four examples of this condition. (The cases are arranged in order of increasing severity of skeletal changes, but the illustrations are arranged in a different order to show particular aspects of the cases side by side. Topographical photographs and radiographs of the vertebral column in Case 4 are followed by low-power histological preparations from the individual cases, then by high-power photomicrographs of the abnormal bone tissue, and finally by pictures of repairing fractures in the individual cases.)

CASE REPORTS

Case 1 (case reported by Maclay, Stokes and Russell 1938)—The patient was a woman aged twenty-nine years, with a history of increasing virilism for three years, associated with depressive mental changes. Glycosuria and hypertension were present but neither signs nor symptoms referable to the skeleton were noted.

At post-mortem examination there was no pituitary adenoma, but hyaline change of the basophil cells was observed. The cortex of each adrenal was hyperplastic. The changes of congestive cardiac failure were present.

The spine showed osteoporosis. The general outline of vertebral bodies and of intervertebral discs was normal (Fig. 8). The cortical bone was thinned and presented many small defects where bone marrow was in contact with either periosteum or intervertebral disc tissue. In the cancellous bone all trabeculae were thinner than normal. The horizontally disposed trabeculae were most severely involved by the rarefaction; many were absent and

the remaining trabeculae were thus predominantly vertical in their arrangement. The vertebral bodies contained considerable amounts of haemopoietic marrow, although histological examination showed an increased number of fat cells (Fig. 11). The slender trabeculae had a normal histological structure, consisting of lamellar bone in which each lamellar system was

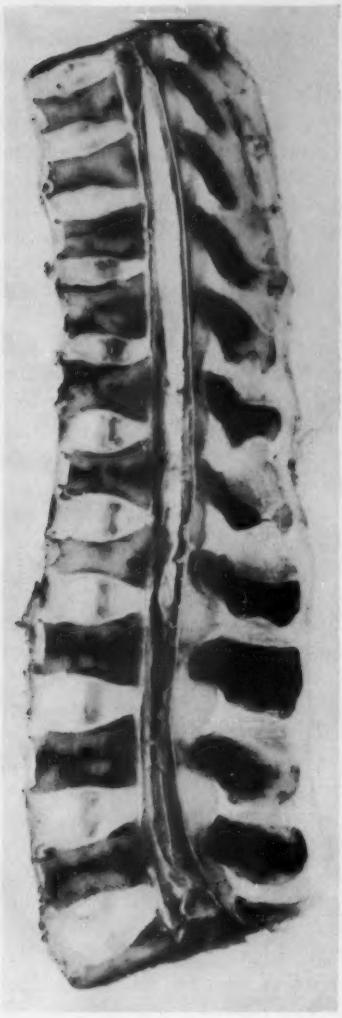

FIG. 1

FIG. 2

Case 4. Figure 1—Photograph of the thoracic and lumbar spine divided longitudinally, showing compression of the anterior parts of the vertebral bodies with expansion of the intervertebral disc tissue. Haemopoietic marrow is replaced by fat in many of the vertebral bodies. Figure 2—A radiograph of the specimen shown in Figure 1, but omitting the upper two vertebrae.

bounded by normal cement lines. Osteocytes and osteoblasts appeared normal. Narrow osteoid borders were seen on the surfaces of a small number of bone trabeculae, and the presence of scattered areas of focal erosion of the trabecular surfaces indicated active, though not excessive, osteoclastic bone resorption.

In histological sections the ribs showed a considerable degree of osteoporosis and contained rather fatty bone marrow. There were some spontaneous fractures and these show developing callus (Figs. 19 to 21). In what appeared to be a very recent fracture there was active bone formation but little vascular reaction (Fig. 20), while in several older fractures the callus tissue was at an unusually quiescent stage despite the incompleteness of bony union.

Sections of the femoral shaft showed slight osteoporosis which took the form of patchy thinning of the cortex, evidence of bone destruction being particularly evident on the endosteal surface. Other bones were not studied.

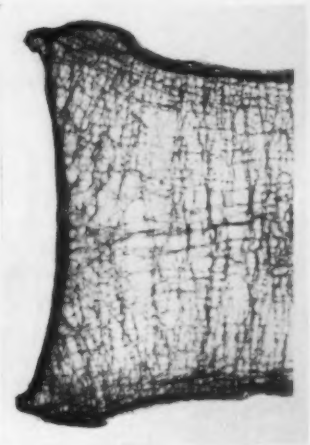

FIG. 3
Radiograph of a slab of bone from a normal vertebra for comparison with Figures 4 to 6. (×2.)

Case 2—The patient was a woman aged thirty-three years. There was a history of amenorrhoea, hirsutes and increase in weight, with evidence of glycosuria, hypertension and renal insufficiency. Radiographs of the skeleton showed osteoporosis of the spine, ribs, and long bones. Several ribs showed fractures with formation of callus, but there was no evidence of collapse of any of the osteoporotic vertebral bodies. Death was due to uraemia and malignant hypertension.

At post-mortem examination the pituitary contained a basophil adenoma 6 millimetres in diameter; microscopically a conspicuous degree of hyaline change affected the remaining basophil cells. The adrenals, which together weighed 16·3 grammes, were of unequal size, the right being partly destroyed by old infarcts. There was great cortical hyperplasia in the left adrenal cortex and in the residual parts of the right cortex.

The spine showed osteoporosis. The general outline of vertebral bodies and of intervertebral discs was normal, but the extent of bony rarefaction was more pronounced than in the previous case (Fig. 9). The cortical bone was very incomplete, particularly on the upper and lower surfaces of the vertebrae, although the general contour of the vertebral bodies was unchanged. In the cancellous bone rarefaction involved both vertical and horizontal trabeculae. There was considerable replacement of haemopoietic marrow by fat cells, which were often arranged along the surfaces of bone trabeculae (Fig. 12). Osteocytes and osteoblasts appeared normal despite diminished osteoblastic activity; the trabeculae were almost completely devoid of osteoid borders. Occasional areas of localised erosion of the trabecular surface indicated continuing, though not excessive, osteoclastic bone resorption (Fig. 12).

In histological preparations the ribs showed marked osteoporosis and contained fatty bone marrow. Material from a spontaneous rib fracture showed developing callus, but although union was incomplete the repairing tissues were at a quiescent stage (Figs. 22 and 23). Thus in Figure 22 the periosteal callus was seen to consist of inactive bone devoid of either osteoid borders or conspicuous osteoblastic activity; Figure 23 showed inactive fibro-cartilaginous callus with little vascularisation or bony replacement.

Sections of femur, pelvis and skull showed slight osteoporosis which took the form of irregular thinning of the cortical bone.

Case 3 (case reported by Lescher and Robb-Smith 1935)—The patient was a woman aged thirty-five years. There was a history of hirsutes, hypertension and glycosuria, together with increasing weight over a number of years. Spinal osteoporosis with collapse of dorsal vertebrae became apparent, and a number of spontaneous rib fractures developed. A large adrenal "cortical cell carcinoma" was present, and the patient died soon after its removal.

FIGS. 4, 5 and 6

Case 4—Radiographs of bone slabs from three of the vertebral bodies shown in Figures 1 and 2. (×2.) Cortical bone is almost completely absent from the osteoporotic vertebrae; the normal cancellous bone is replaced by an incomplete network of irregular trabeculae, which are often greatly reduced in thickness. The anterior and upper parts of two of the vertebral bodies show collapse of the osteoporotic bone structure.

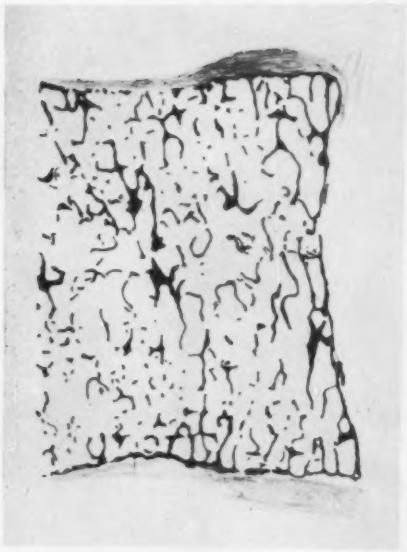

FIG. 7

FIG. 8

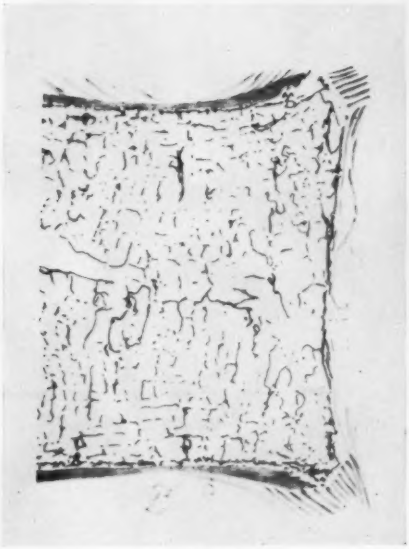

FIG. 9

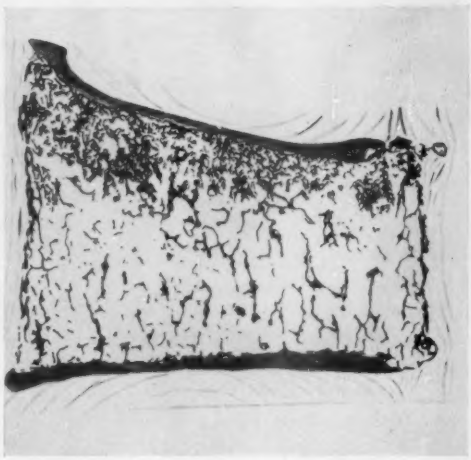

FIG. 10

Outline drawings of the bony structure of vertebral bodies as seen in histological sections. (×2.) Figure 7—Normal structure. Figure 8 (Case 1) and Figure 9 (Case 2) show a diminution in thickness of bone trabeculae. Figure 10 (Case 3)—There is, in addition to this change, compression of the upper part of the vertebral body. Numerous slender newly formed bone trabeculae can be seen in the area of collapse.

At post-mortem examination no metastases were found. The pituitary showed some increase in basophil cells; at one point these formed a small collection 0·3 millimetre in diameter. Extensive hyaline changes in the basophils in this case were reported by Crooke (1935).

The spine showed osteoporosis with collapse of a number of vertebral bodies (Fig. 10). In addition to the general rarefaction of the cortical and cancellous bone, it was evident that the upper surface of the vertebral body illustrated had been displaced downwards with disorganisation of the adjacent tissues. The bone of the lower part of the body was not affected by this compression and displacement. In vertebral bodies which were not collapsed the slender bone trabeculae were almost completely devoid of osteoid borders; they showed evidence of continuing, though not excessive, osteoclastic bone resorption. There was considerable replacement of haemopoietic marrow by fat cells. In the collapsed vertebrae, many individual

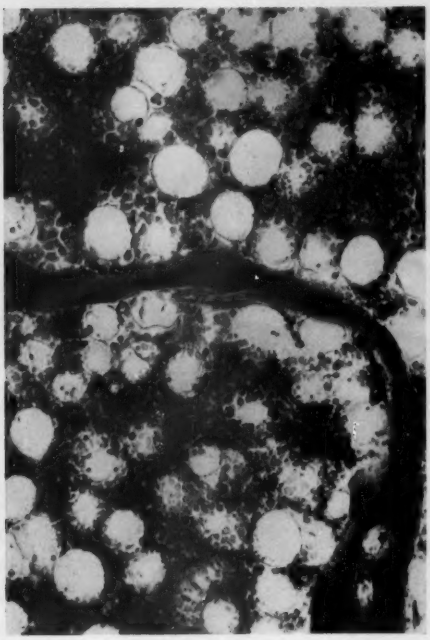

FIG. 11

Case 1—A slender bone trabecula from the central part of a vertebral body showing a narrow osteoid border on its upper surface. (×75.) The opposite surface of the trabecula presents an eroded appearance.

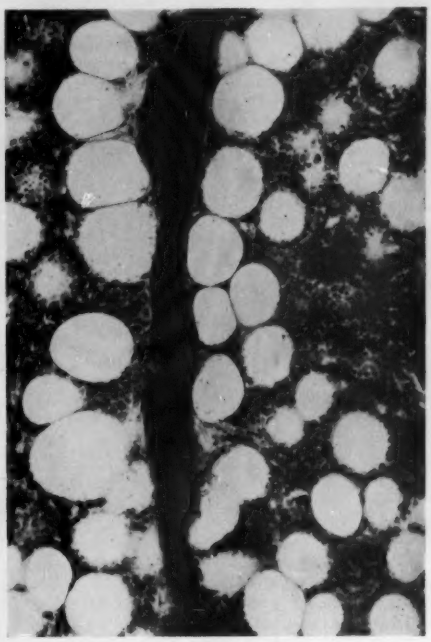

FIG. 12

Case 2—A slender bone trabecula from the central part of a vertebral body. (×75.) Occasional eroded bays are present on the bone surface, indicating some previous osteoclastic resorption.

bone trabeculae were fractured and displaced (Figs. 16 and 17). In some areas this damage was associated with only minimal tissue reaction, and had not led to any reparative bony changes. In others, seen at a later stage, the displaced trabeculae were surrounded by irregular masses of newly formed non-lamellar bone.

In histological preparations the ribs showed marked osteoporosis and contained fatty marrow. Sections from a spontaneous rib fracture showed some bony union both endosteally and periosteally (Figs. 24 to 26), but the periosteal callus included a considerable amount of abnormal degenerated cartilage and quiescent fibrocartilage with little evidence of vascularisation and bony replacement. As in the previous cases, there was little activity of periosteal bone formation at the peripheral parts of the callus.

No other bones were studied in this case.

Case 4—The patient was a girl aged sixteen years. There was a history of amenorrhoea and of progressive obesity and hirsutes for five years. Three years after the onset of symptoms, severe lumbar pain directed attention to the spine. The patient was admitted to hospital, and spinal osteoporosis with collapse of several vertebral bodies was found. Diabetes and hypertension were present at this stage. Surgical exploration of the adrenals showed symmetrical cortical hyperplasia; the patient's condition temporarily improved after the removal of the left adrenal.

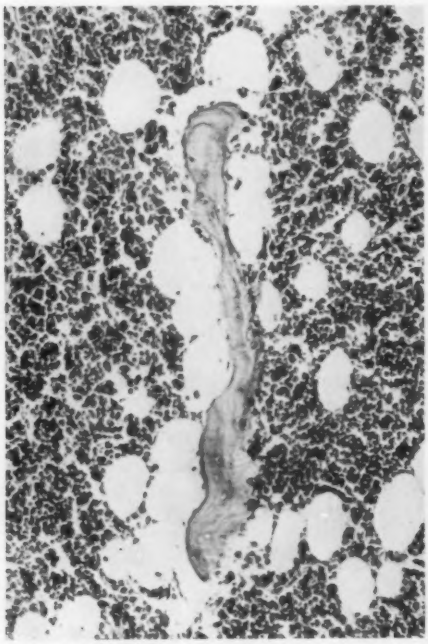

FIG. 13

Case 4—A slender bone trabecula from the central part of one of the vertebral bodies. (×75.) Note the increased number of fat cells present in the marrow; many of these fat cells are in contact with the surface of the bone trabecula.

At post-mortem examination the pituitary contained a basophil adenoma; microscopically there was a conspicuous degree of hyaline change affecting the remaining basophil cells. Recent haemorrhage had occurred both in the tumour and in the surrounding pituitary tissue. There was some degree of hyperplasia of the cortical tissue of the remaining adrenal.

The spine showed a marked degree of osteoporosis with a varying degree of collapse of a number of vertebral bodies in the lower dorsal and lumbar region (Figs. 1 and 2). The contour of the vertebral bodies was changed, the concavity of the upper and lower surfaces being greatly exaggerated with a corresponding "ballooning" of the intervertebral discs. In collapsed vertebrae it was the anterior part of the body which was affected. The extreme rarefaction of cortical and cancellous bone became evident when Figures 4 to 6—radiographs of bone slabs from vertebral bodies in this case—were compared with Figure 3, which showed a similar preparation from a normal vertebral body. Some areas of the abnormal vertebral bodies were completely devoid of bone trabeculae, while those trabeculae present in other areas were abnormally slender. They were often angulated, and irregular and crenated in outline. In all the vertebrae there was considerable replacement of haemopoietic marrow by fat, this being most apparent in the areas where bony rarefaction and collapse were most marked (Fig. 1). The surfaces of the slender bone trabeculae of the osteoporotic vertebral bodies were completely devoid of osteoid borders (Fig. 13). Although such osteoblasts as were present were histologically normal, their number was very much diminished, and their total bone-forming activity was clearly reduced. There was evidence of continuing, but not excessive, osteoclastic bone resorption, although osteoclasts themselves were rare. The cortical bone in the vertebral bodies, as well as being greatly thinned (Fig. 14), presented many small defects (Fig. 15) where the bone marrow was in contact with either periosteum or intervertebral disc tissue. Despite the changed contour of the upper and lower surfaces of vertebral bodies, the line of junction between the bone and the adjacent intervertebral disc tissue was smooth. Bone formation extended into the cartilage of the intervertebral disc in some areas, as was evident in the radiographs of bone slabs shown in Figures 4 to 6, but the general contour of the bone surface is not interrupted.

In the collapsed vertebrae there were numerous fractures of individual trabeculae. In most areas the reaction of the adjacent tissue to recent trabecular fractures was minimal,

although in one vertebra the displaced trabeculae in the collapsed area were surrounded by vascular granulation tissue and proliferating osteoblasts. Despite the minimal reaction, the fractured and displaced trabeculae eventually became bound together into irregular masses of bone (Fig. 18). The structural changes in these vertebrae (see for instance Figures 2, 4, 5 and 6) indicate that " collapse " of the osteoporotic bone structure may take place quietly and continuously, but that this process can sometimes be interrupted by a sudden and more complete compression fracture.

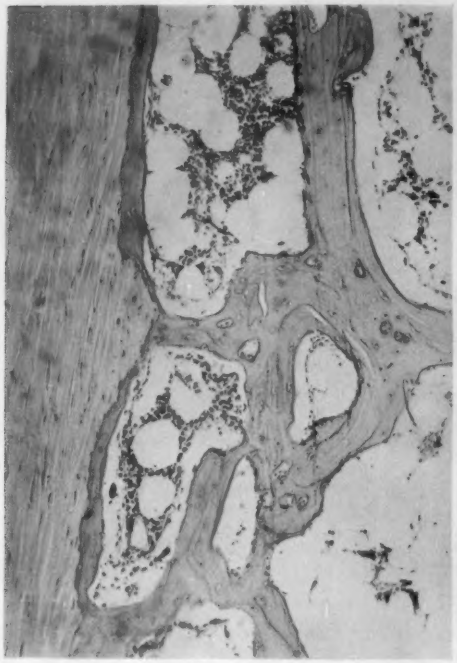

FIG. 14

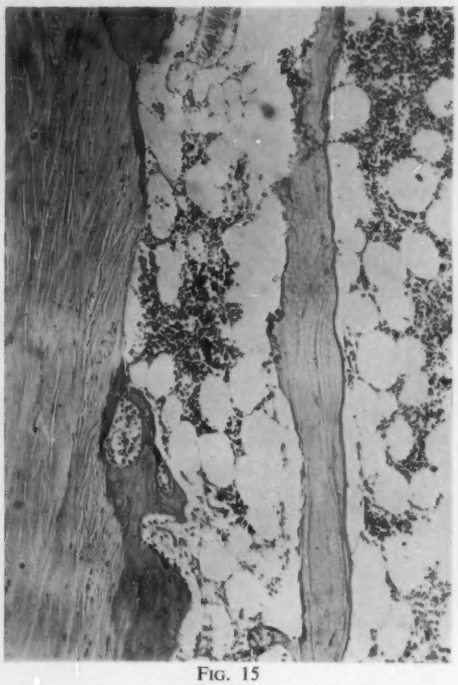

FIG. 15

Case 4—Areas from the rarefied cortical bone of the anterior wall of a vertebral body, showing extreme narrowing of the bone and the contact between bone marrow and periosteal fibrous tissue in some areas. (×75.)

Sections of the ribs showed osteoporosis, both the cortical bone and the medullary trabeculae being thinner than normal. Sections of femoral shaft showed slight osteoporosis in the form of patchy thinning of the cortical bone. Sections of the vault of the skull did not show any histological abnormality.

DISCUSSION

In 1920 Mooser included a detailed description of bone changes in his account of the post-mortem findings in a case of " obesity with osteoporosis " which Cushing recognised as comparable with his own cases. Mooser showed that the bone change was indeed osteoporosis, described the collapse of vertebral bodies without evidence of acute fracture, and noted that evidence of exaggerated osteoclastic bone destruction was lacking. The vertebrae in the case he described showed unusual herniations of bone marrow into adjacent degenerated and calcified intervertebral disc tissue, and also herniations of disc tissue into the vertebral bone marrow that would now be described as Schmorl's nodes. In 1933 Rutishauser noted and described the osteoporosis present in two cases of Cushing's syndrome coming to post-mortem examination, each case being associated with a pituitary adenoma. More recently Follis (1951*a*) has described bone changes in five cases of Cushing's syndrome studied

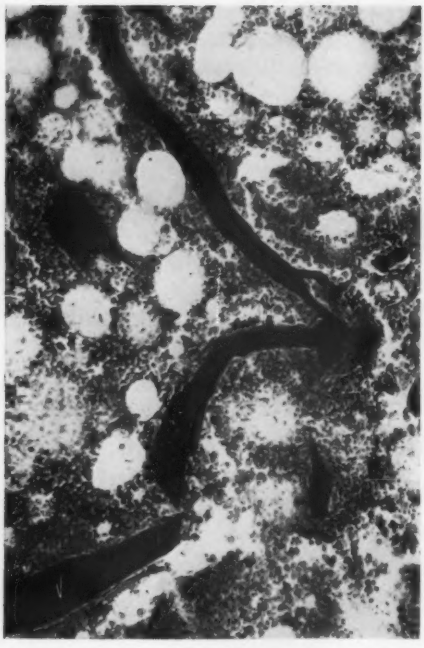

FIG. 16

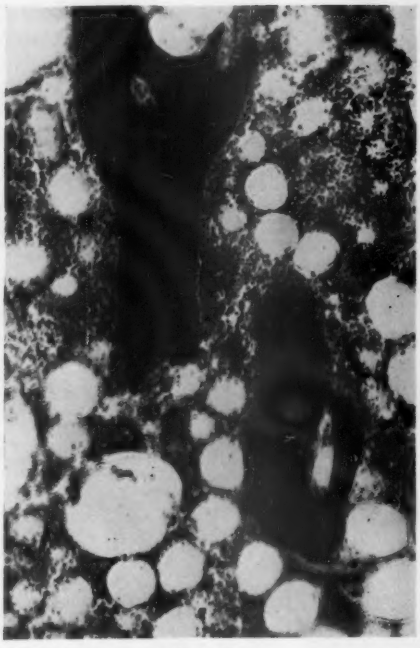

FIG. 17

Case 3. Figure 16—Fractured bone trabecula at the margin of the area of collapse in the vertebral body shown in Figure 10. (×75.) Reaction of the adjacent tissues is minimal. Figure 17—Another fractured bone trabecula from the collapsed area of the vertebral body shown in Figure 10. (×75.) Some displacement has occurred, but again the reaction of the adjacent tissues is minimal.

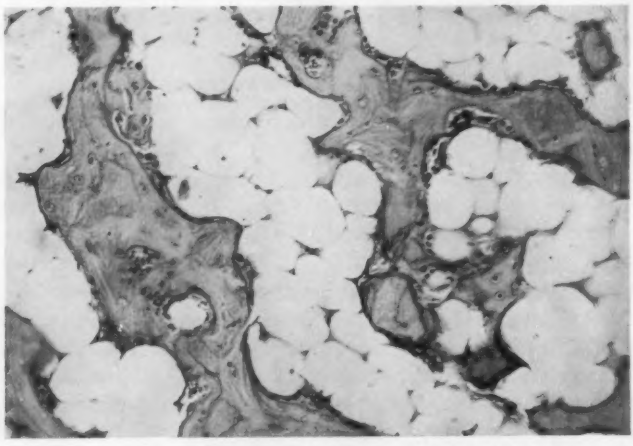

FIG. 18

Case 4—Bone trabeculae in the collapsed area of the vertebral body shown in Figure 5. (×75.) The appearances suggest that slender trabeculae in the collapsed region have become impacted, and have been surrounded by newly formed bone to produce thicker structures.

FIG. 19

Case 1—A histological preparation of a rib showing two recent and incompletely united spontaneous fractures. The larger one is centrally placed and is surrounded by periosteal callus; the other is toward the bottom of the figure and shows neither displacement of the bone nor periosteal callus.

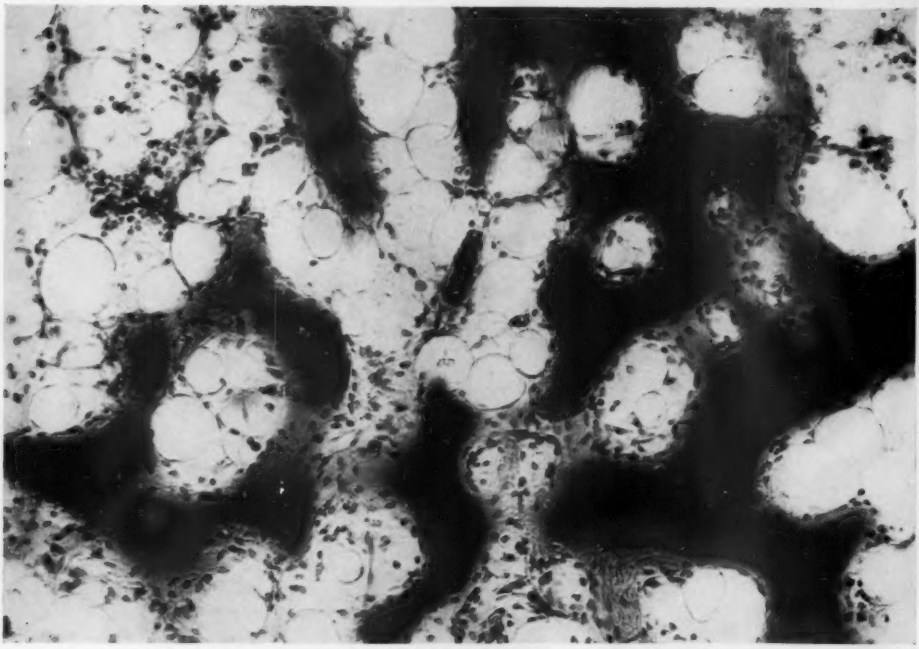

FIG. 20

Case 1—Showing active osteoblastic bone formation in the endosteal callus of the smaller and very recent fracture in Figure 19. (×160.)

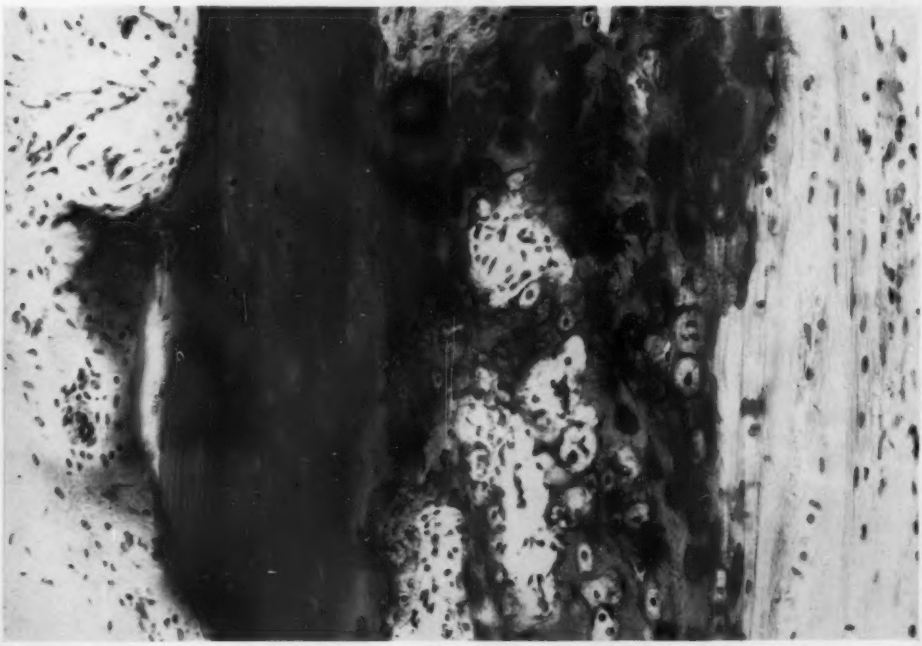

FIG. 21

Case 1—Showing less active periosteal callus from the larger and older fracture in Figure 19. (×160.)

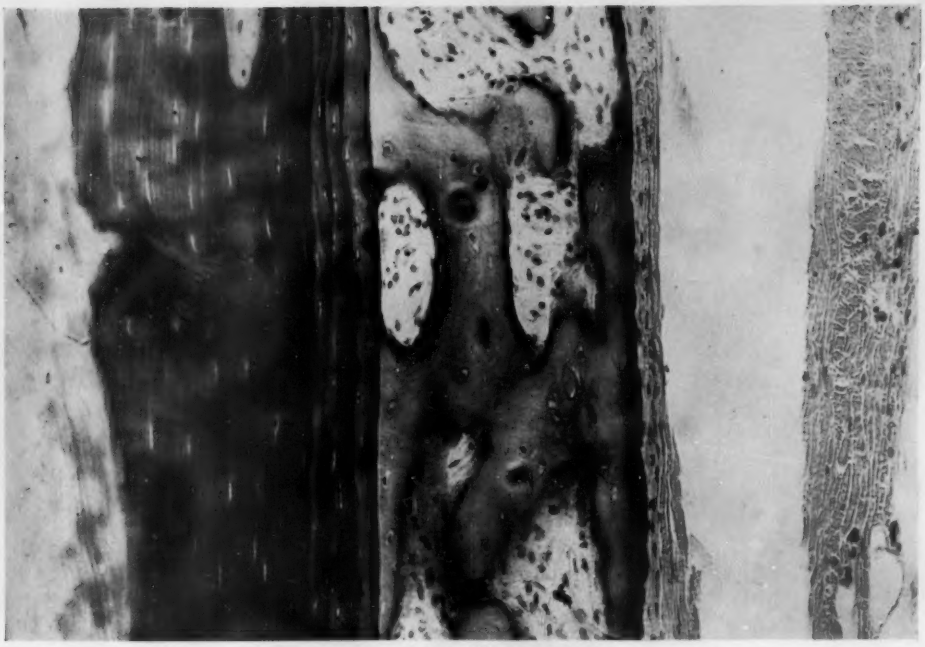

FIG. 22
Case 2—Inactive periosteal callus from spontaneous fracture of rib. (×160.) Few osteoblasts are present on the surface of the newly formed bone.

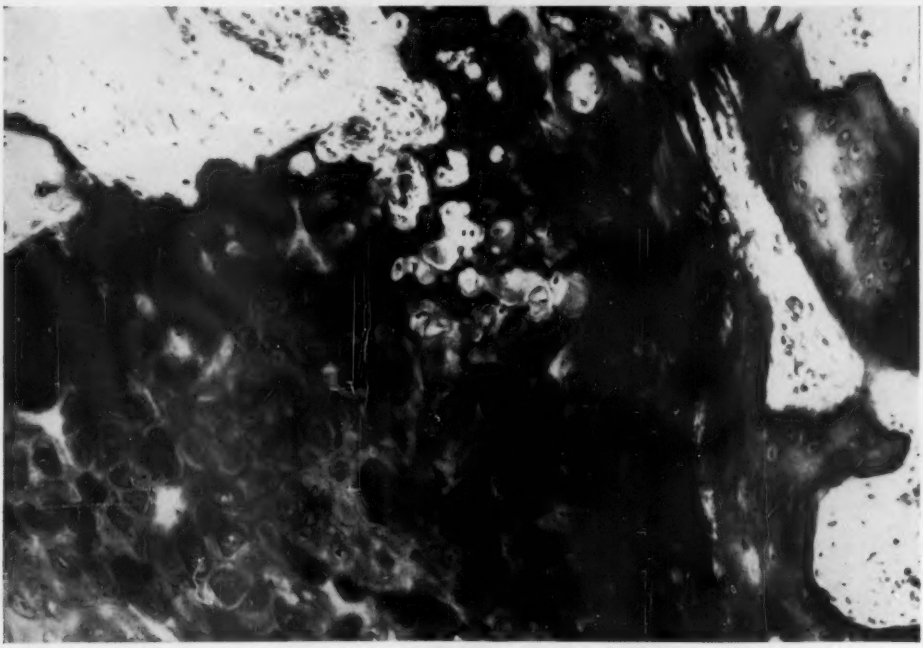

FIG. 23
Case 2—Inactive periosteal callus from spontaneous fracture of rib. (×160.) The field shown consists of unusual fibrocartilaginous callus showing only minimal vascularisation.

FIG. 24

Case 3—A histological preparation of a rib showing a spontaneous fracture. (×4.) The progress of union is seen at a later stage than in Figures 19 to 21. Both periosteal and endosteal bony callus are uniting the bone ends.

FIG. 25

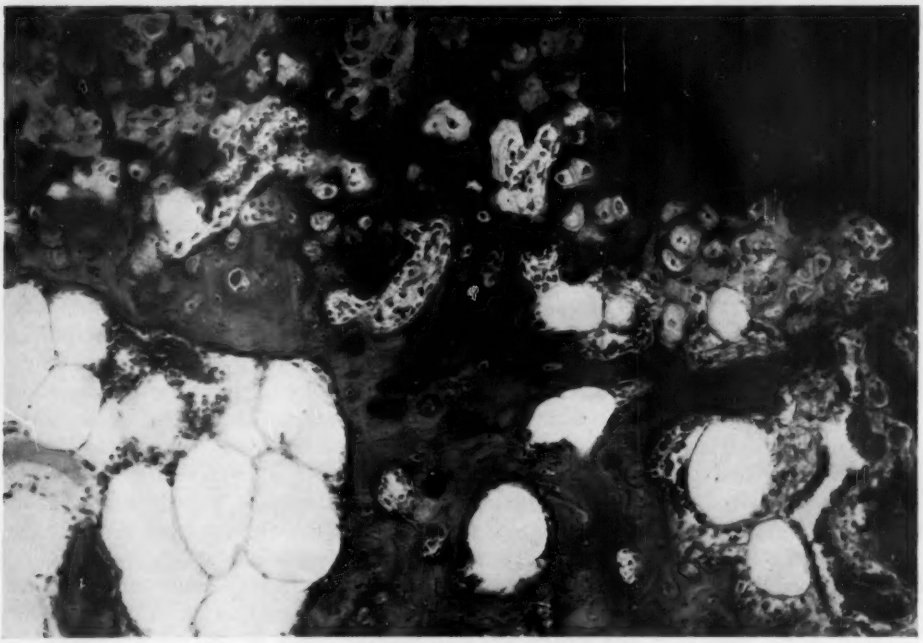

FIG. 26

Case 3—Further preparations of the specimen seen in Figure 24. (×160.) Unusual degenerate and fibro-cartilaginous callus is seen, with little vascularisation and bony replacement.

at post-mortem. In four adult cases he found osteoporosis. Osteoid borders were occasionally seen, but osteoclasts were not observed. In his fifth case Follis recorded the changes in growing epiphysial cartilage in a " Cushing-like " condition in an infant of fifteen months. These took the form of growth arrest and disorganisation of cartilage tissue.

The changes described in the vertebral bodies in the present series of cases are substantially those of the accounts mentioned above. The abnormalities of fracture repair noted do not appear to have been recorded previously. Despite the bony collapse present in some cases, the Schmorl's nodes observed by Mooser were not present in the vertebrae examined in the present series.

In contrast to the osteoporosis of hyperparathyroidism (Hunter and Turnbull 1931, Jaffe 1940) or of hyperthyroidism (Follis 1953) there is no evidence that increased osteoclastic destruction is the factor responsible for the changed bone structure in the osteoporosis of Cushing's syndrome. Indeed, in the material that he studied, Follis was not able to find any osteoclasts at all! The diminished number of osteoblasts on the bone surfaces in the most severely osteoporotic cases, and the scarcity of osteoid borders, both indicate, however, that interference with osteoblastic bone formation is the important factor in the production of skeletal changes. This histological abnormality is, in fact, what might be expected from Albright's hypothesis that the effect of the abnormal or excessive adrenal hormones in Cushing's syndrome is to inhibit the synthetic metabolic activities of tissues. The maintenance of normal bone structure depends, ultimately, on a balance between osteoblastic bone formation and osteoclastic bone destruction. Any persistent disturbance of this balance in favour of destruction, whether an exaggeration of osteoclastic bone destruction or an inhibition of osteoblastic bone formation, would give rise to skeletal rarefaction. In Cushing's syndrome the second of these alternatives appears to be the one concerned.

While changes in the shafts of long bones are not as evident as those in vertebral bodies in Cushing's syndrome, it would be of great interest to examine bone from such situations, using the techniques of microradiography developed by Engström and Amprino (1950). Even in areas where osteoporosis was not evident, such investigations might be expected to show interference with the normally continuous process of formation and progressive calcification of bone in the Haversian systems of compact bone.

It is of interest to compare the skeletal changes in Cushing's syndrome in man with those produced experimentally in animals by the administration of either pituitary adrenocorticotrophic hormone or cortisone. Thus Becks *et al.* (1944) described inhibition of proliferation of cartilage cells and of osteoblasts in the region of the epiphysial plate in the bones of young rats following the administration of A.C.T.H. The effect of cortisone and of 17-hydroxycorticosterone-21-acetate on the growing bones of the rat has been described by Follis (1951*b*, 1952), while Sissons and Hadfield (1955) have studied the effect of cortisone on the growing bones of rats and rabbits. In the rabbit, cortisone inhibits the proliferation of the cartilage cells of the epiphysial plate, with reduction in the rate of longitudinal bone growth, even down to complete cessation. Osteoclastic activity is unimpaired, and progressive resorption of metaphysial trabeculae results from the inhibition of osteoblastic bone formation. Thus the skeletal position is exactly comparable with what is found in Cushing's syndrome. In the rat, cartilage proliferation is inhibited and longitudinal growth is diminished, but the effect on bone itself is inconstant. Sometimes resorption of metaphysial trabeculae is seen; at other times the balance between osteoblastic and osteoclastic activity is changed in the reverse direction and metaphysial trabeculae of abnormal density are produced.

The diminution in the reparative processes of fracture repair described in the present paper corresponds with the results of experimental work with cortisone. In rabbits, for example, Blunt *et al.* (1950) and Sissons and Hadfield (1951) have shown that administration of cortisone inhibits both cartilage proliferation and osteoblastic bone formation in the developing callus of experimental fractures. In the experimental fractures in cortisone-treated

animals, moreover, the latter authors have described the same type of quiescent and degenerate cartilaginous tissue, showing little or no evidence of vascularisation, that is found in Cases 1, 2 and 3 of the present paper.

SUMMARY

1. The bone changes in four autopsied cases of Cushing's syndrome are described. The changes take the form of osteoporosis, which is most marked in the spine and the ribs.

2. The osteoporosis results from impaired osteoblastic bone formation in the presence of a normal degree of osteoclastic bone resorption.

3. Histological abnormalities of fracture callus in Cushing's syndrome indicate interference with the proliferation of osteoblasts and cartilage cells and with the formation of new tissue by these cells.

4. The bone changes in Cushing's syndrome are comparable with those produced in experimental animals by the administration of A.C.T.H. or cortisone.

I am greatly indebted to Professor Dorothy Russell for access to pathological material and information from Cases 1, 2 and 3, to Dr E. F. Scowen and to Professor J. W. S. Blacklock for similar help with Case 4, and to Dr M. H. Jupe for information regarding the radiological findings in Case 2. The radiographic part of the work was supported by the James Picker Foundation on the recommendation of the Committee on Radiology, National Academy of Sciences, the National Research Council of the United States of America.

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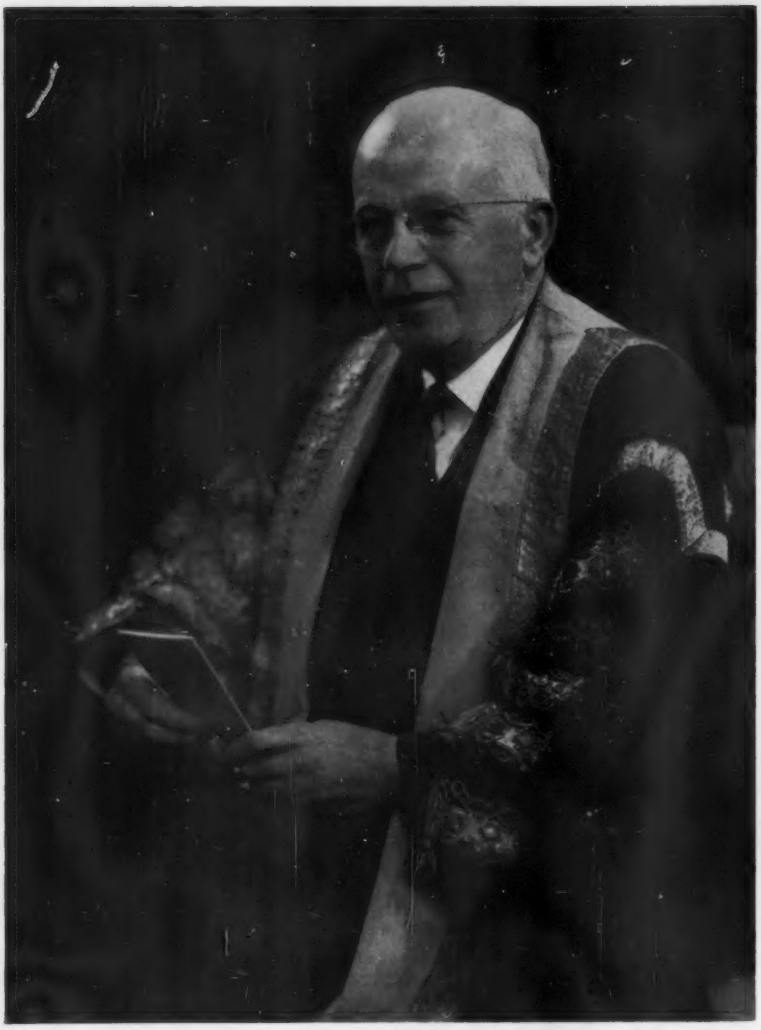

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CHAIRMAN OF THE BRITISH EDITORIAL BOARD

We are proud to know that Walter Mercer, chairman of the British Editorial Board of the *Journal of Bone and Joint Surgery*, has recently been recognised for his great work and achievement by two signal honours. This month he received the accolade of Knighthood from Her Majesty Queen Elizabeth the Queen Mother. As Her Majesty laid the sword on his shoulder he became Sir Walter Mercer; and his wife, who has always been the perfect partner as well as hostess and friend to all his colleagues and students, is now Lady Mercer.

Then in recent weeks the President and Council of the Royal College of Surgeons of England conferred upon him their honorary fellowship which is guarded so jealously that no more than forty surgeons throughout the world may hold it. There was only one vacancy in the list of distinguished honorary fellows of this College and yet Sir Walter Mercer was elected promptly and with enthusiastic unanimity.

Why was this so? First because he has been so great a general surgeon, ready with expert skill to apply himself to the surgical problems of the upper abdomen, the urogenital tract, the lungs and heart, the brain, the spinal cord, or indeed any area where the technique of surgery has advanced so fast that most of us have been satisfied to concentrate upon one region alone—but not so for him. He can nail a fractured neck of the femur more quickly than others; but he can also divide a stenosed mitral valve, resect a lung, or remove a brain tumour.

It should be noted that he has not dibbled and dabbled in these fields of surgery all at the same time. For some years he concentrated on abdominal surgery, then on chest surgery, then on neurosurgery, and at last by reason of pressure from the University of Edinburgh accepted the Chair of Orthopaedic Surgery to which his practice and teaching is now devoted.

Five years ago he was elected President of the Royal College of Surgeons of Edinburgh. His contribution to this noble and ancient college is perhaps greater than that of any president before. He looked back to old traditions, for example restoring the duties of the chaplain who adds such dignity and honour to the proceedings of his College; but he also looked forward to the future, and made certain that the Edinburgh College should not be a simple forum of examination but also an academic centre of training in basic medical sciences as well as in the practice of surgery.

It is recorded that his recreations are curling, shooting, golfing, fishing and the history of stamps. Curling " just comes natural " to a Scotsman so we need say no more about it. In shooting it may be that his alignment of a gun is no longer as good as his alignment of a bone. He is a golfing-widower because his wife's handicap is less than his. He is also a fishing-widower and usually serves as ghillie to his more expert wife. She however is a stamp-widow because his knowledge of philately is so great. The amazing thing about Walter Mercer is his perennial youth. He was kicking a football at George Watson's College in Edinburgh before many of us were born—and yet he seems younger than ever.

We know his kindly and friendly hospitality. We are met by Sir Walter himself at six or seven o'clock in the morning on the station platform when wearily turning out from the overnight train from the South—and yet the warmth of his reception is such that at least one of us who has enjoyed early morning baths and breakfasts in his home is known to his housekeeper as " the gentleman who sings in the bath."

For all these reasons, and for many others, members of the British Editorial Board of this Journal want to say how proud they are of their Chairman. REGINALD WATSON-JONES.

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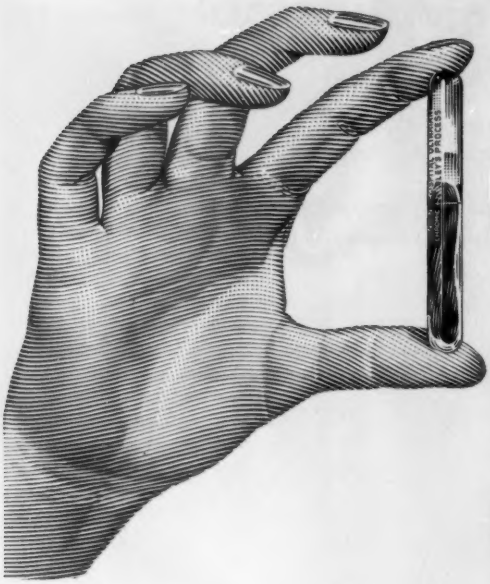

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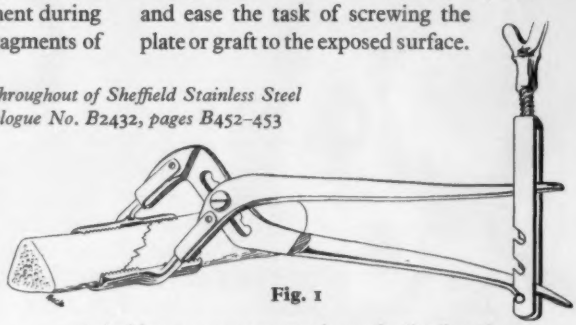

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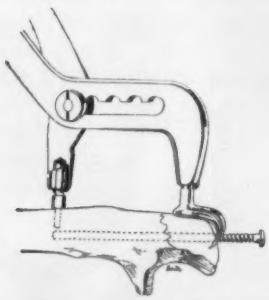

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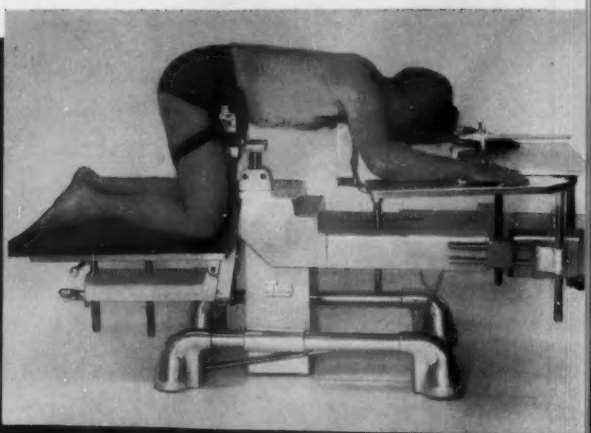

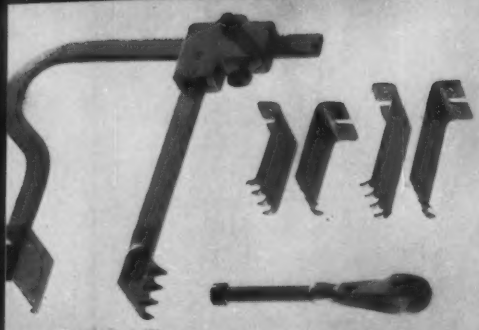

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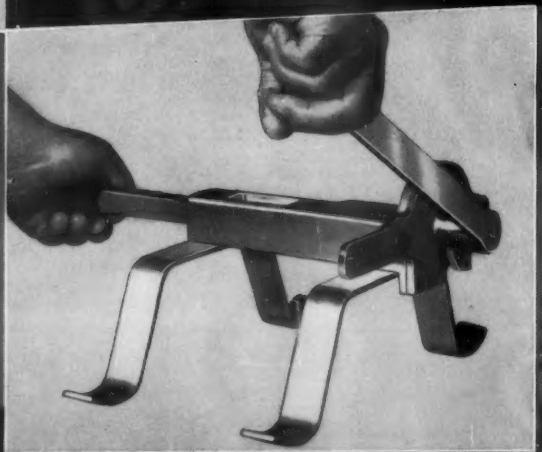

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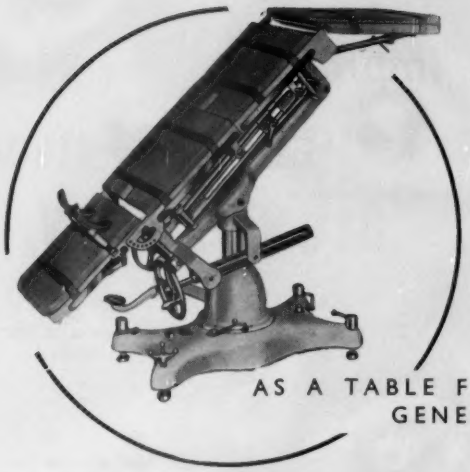

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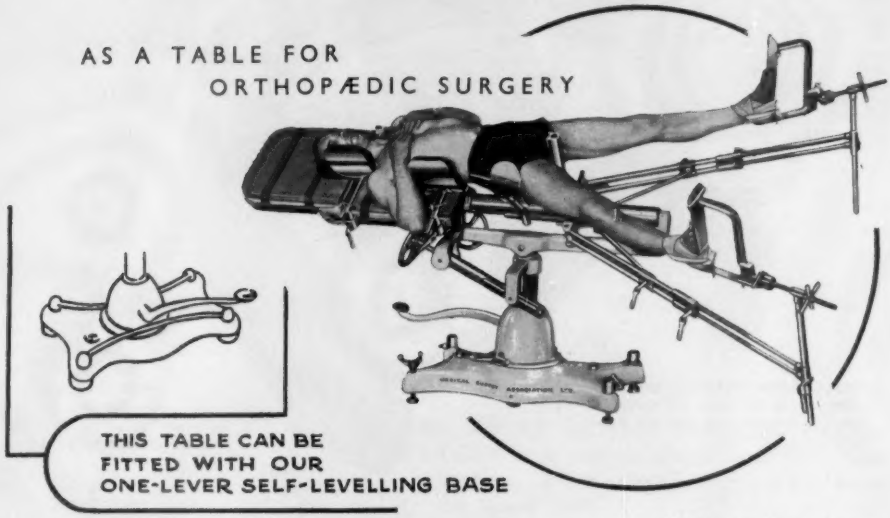

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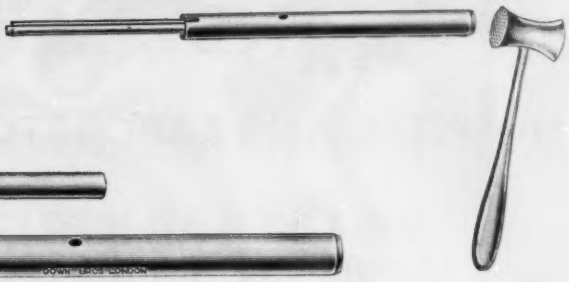

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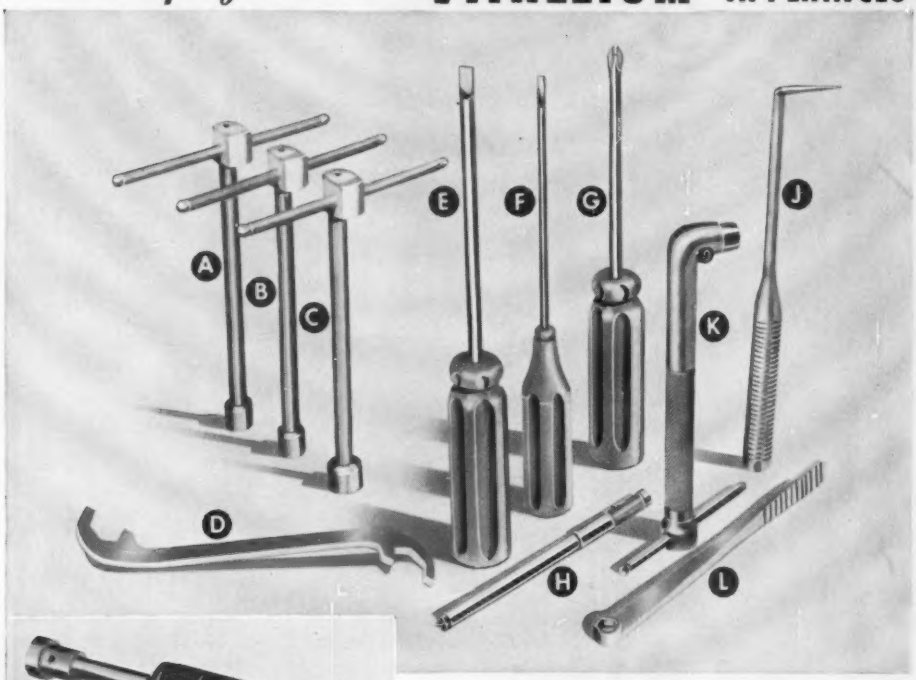

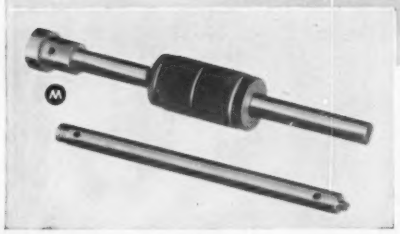

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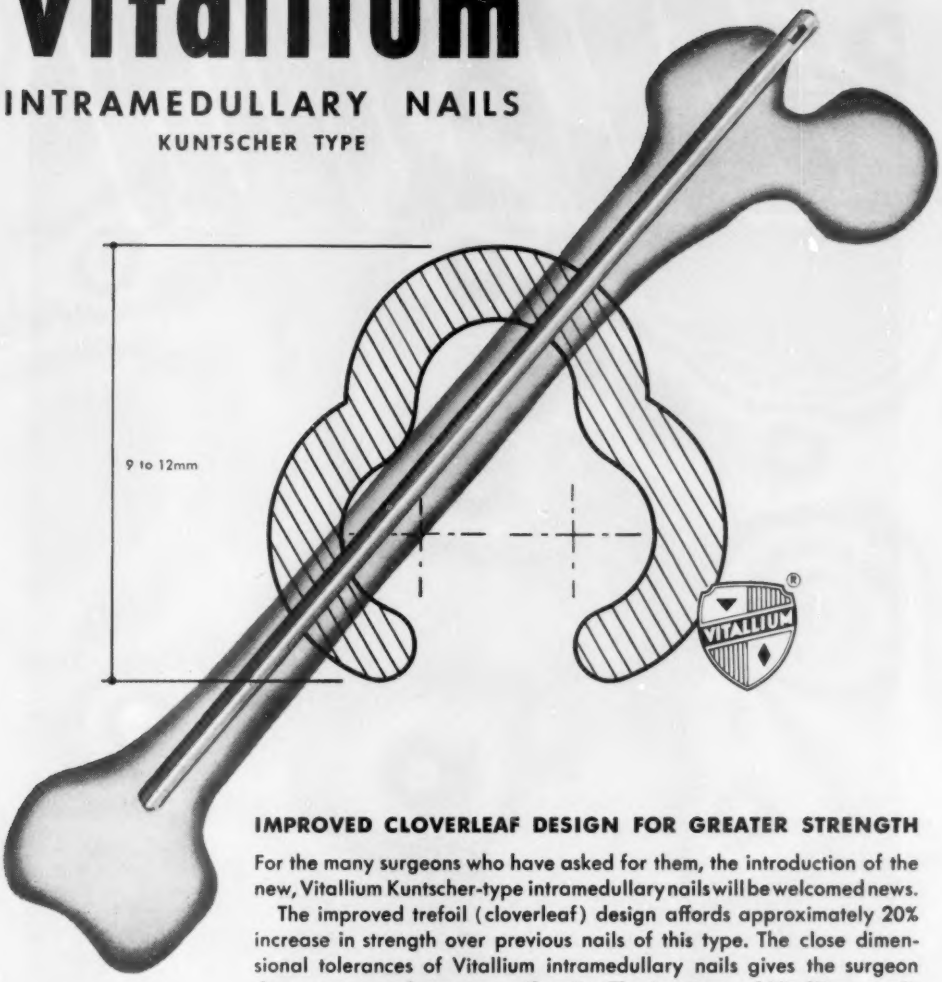

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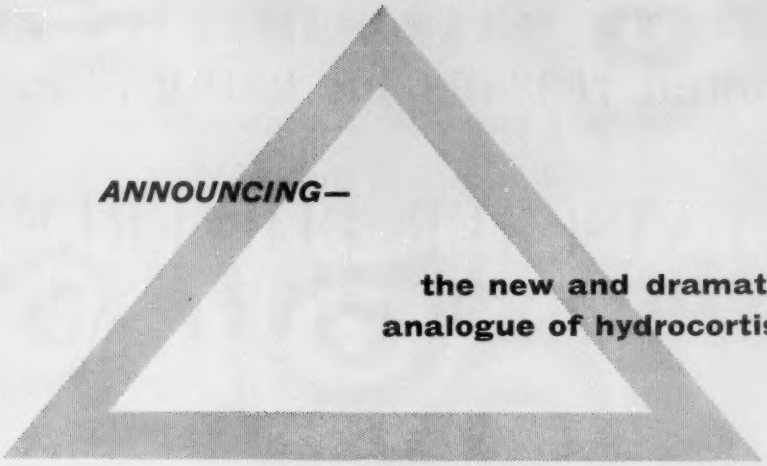

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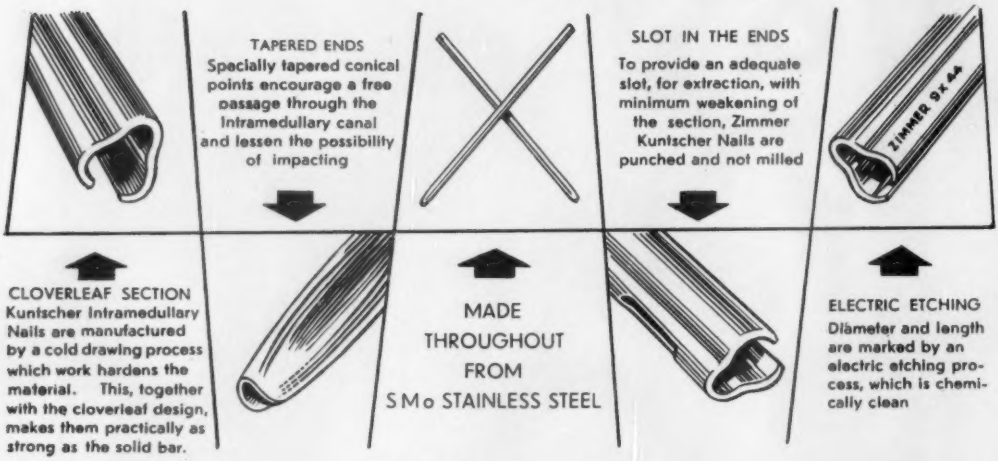
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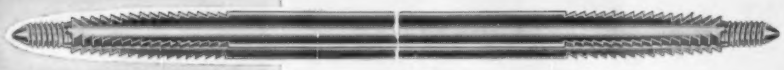

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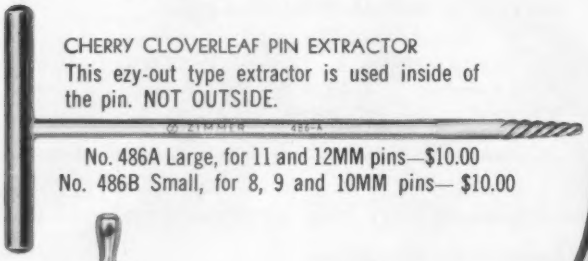

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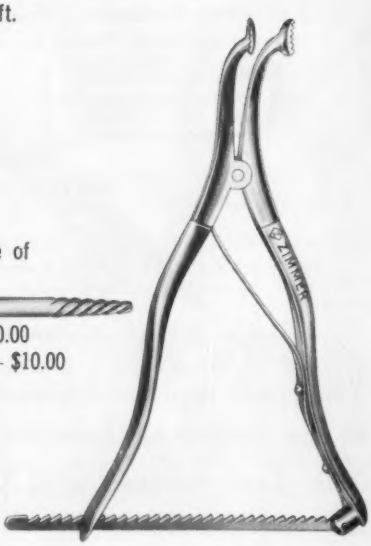

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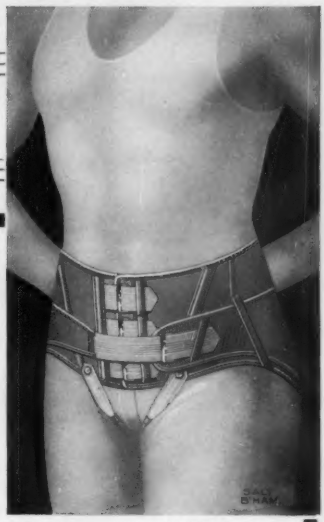

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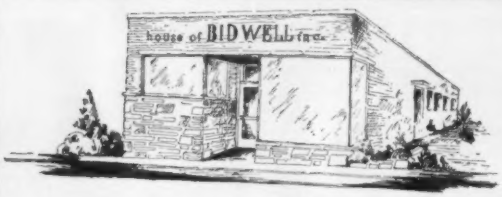

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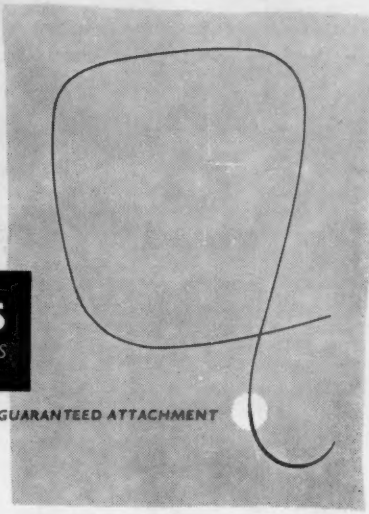

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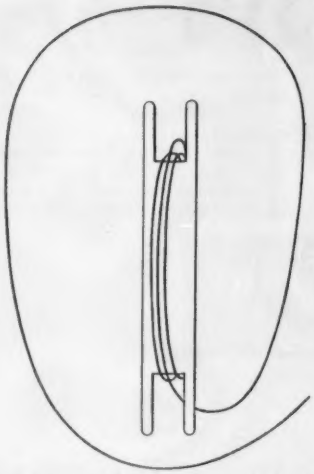

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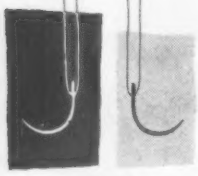

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